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THE UPWARD MOVEMENT OF THE EYES.[1]

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NEW points of view evolve with the growth of knowledge. Thus there has been a natural tendency of recent years, which becomes increasingly more apparent as information regarding normal and pathological structure accumulates, to regard neurological problems in relation to conceptions of function rather than from the strictly anatomical standpoint. The genius of two former presidents of this Society—Dr. Hughlings Jackson and Sir Charles Sherrington—has been largely responsible for the attainment of this outlook. In like manner our attention will no doubt be focused more and more upon the genesis of disease rather than upon its results, as represented by the pathological changes in the tissues.

The clinician is continually engaged from his earliest days in acquiring a practical knowledge of disease. Most of us pass through a stage during which the rarities and the unusual attract our attention and appeal to our enthusiasm, while at a later period a closer study of the commoner disorders and phenomena, with particular reference to the application of general principles, engenders a fresh interest. With increasing experience one's outlook widens, one learns to look at things from different points of view, and one is more and more conscious of personal limitations, and impressed by the limitations of knowledge and by the great field for clinical observation which lies before us.

I have selected as the theme for these remarks "the upward movement of the eyes." My purpose is to indicate some of the many questions which arise when we regard an apparently simple phenomenon

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from a variety of standpoints. The title I have chosen is perhaps misleading, for it is not my intention to attempt an exhaustive disquisition. Again the choice of subject may appear to be presumptuous, for I do not flatter myself that anything I have to say is new to you.

Some weeks ago, just as the light was failing, I was struck by the behaviour of an owl perched upon an isolated branch. The small birds around him were twittering in alarm, and from time to time the owl turned his head suddenly, first to one side then to the other. The observation was new to me, although probably familiar to some of you, for my children tell me that they have often watched an old owl in the Zoological Gardens and noticed these sudden movements of the head. A simple observation may serve to arouse a train of thought. Presumably the movements of the owl were determined by the sounds in his vicinity, and presumably he turned his head in order to visualize the better his surroundings. Why is it that in man we never see these sudden movements of the head, for the eyes of man like those of the owl are set far forward and the owl's vision, as is the case in man, must be binocular? I recalled the observations of Wilfred Harris [1] who noted that birds have little if any power of movement of the eyes, and that in the birds, including the owl, there is a complete decussation of the visual fibres at the chiasma. The owl is obliged to move his head since he cannot otherwise fix his eyes upon his prey. Such sudden movements of the kind must make his presence more evident, yet this is not perhaps a matter of material moment since the owl is a bird of the night and in this respect peculiar.

The co-ordinated movements of the head and eyes in man are obviously regulated by a mechanism of extreme beauty and delicacy. We are constantly moving the head and eyes for purposes of vision, the object to which our attention is directed providing the stimulus which determines the movement called for. The visual movement mechanism in man is determined by the demands of stereoscopic vision, and the importance of these co-ordinated movements is apparent when we recall that they constitute a precursor to the acquisition of specialized purposive acts, even though such acts may subsequently become more or less automatic by repetition.

Then, too, it is interesting to note that the relative degree to which we bring into play the movements of the head and of the eyes respectively in relation to visual fixation must be fairly constant under normal conditions, for, although both sets of movements can be utilized independently for visual purposes, pronounced rotation of the head to

one side without an associated deviation of the eyes, or pronounced deviation of the eyes to one side, the head remaining fixed, are movements so unusual that they at once arrest attention. The actor is well aware of this and he exaggerates those independent movements of the head and eyes to which a conventional meaning has come to be attached for purposes of emphasizing his emotional expressions.

Further, the co-ordinated movements of the head and eyes afford a very beautiful illustration of the dictum, which we associate with the name of Dr. Hughlings Jackson, that movements not muscles are represented in the cortex. Indeed the contraction of the homolateral sternomastoid in association with deviation of the eyes to the opposite side, as seen both on voluntary movement and in the cortical fit, is perhaps the most striking instance of the preponderating participation of a homolateral muscle in a co-ordinated movement.

The acquisition of co-ordination is a subject of entrancing interest, raising as it does questions of functional adaptation. Is it not a remarkable fact that the new-born chick should walk about and pick up its food as soon as it emerges from the egg, whereas the young nestling is at first so helpless? Co-ordinated movements are for the most part acquired and not innate and, speaking generally, the more specialized a co-ordinated movement the later its development. This certainly applies to the more highly specialized movements of man.

All of us have watched with interest the inco-ordinate movements of the young infant. The eyeballs are seen to roll in various directions, and they tend to move to some extent independently of one another with the result that the child often squints. Preyer [2], who has made this matter the subject of special study, writes: "Some new-born children actually make associated, co-ordinated movements of the eyes several times on the first day: others do not, but I never found in any child co-ordinated movements exclusively . . . . on the whole, I have found that, in the newly-born, one eye very often moves independently of the other, and the turnings of the head take place in a direction opposite to that in which the eyes move. The unintentional character of both movements is plainly recognizable, and the combination of the two is, at the beginning of life, accidental. The turning of both eyes to left and right, also, which is established on the first day, takes on the appearance of accident, coming in as one among all possible movements." The co-ordinated movement of the eyes determines the ability to fixate, and fixation of the eyes is a voluntary movement determined by the will. Gradually the inco-ordinate movements become less noticeable

until, by the end of the third month or thereabouts, they are no longer observed. The innervation of the eye muscle offers, as Sir George Berry [3] truly remarks, "one of the most beautiful and most easily studied examples of the development of a habit." But it is further of interest to note that whereas, according to Preyer, the ability to follow a moving object with the eyes is not usually noticeable before the close of the third month, the ability to turn the head towards a bright light, so that this can produce an image on the retina, is often present on the day of birth. We see here an illustration of the principle that more specialized movements are acquired at a later date than those which are less specialized.

It is a remarkable fact that once the habit of co-ordination of the eye movements has developed in response to visual fixation, the eye muscles cannot be individually activated. Most of us can probably cite exceptional cases in which an individual was able to contract an isolated muscle apart from a co-ordinated movement in which it regularly takes part, but I know of no instance in which, the ocular muscles being intact, it was possible voluntarily to move one eye independently of the other. The call for co-ordination in relation to vision would seem to be so powerful, and the cortical representation of the co-ordinated movements consequently so indelibly fixed as such, that this is an impossibility. It would indeed appear that, if a habit movement is developed in relation to an essential function, no isolated contraction of a muscle concerned can be brought about should this in any way interfere with the specific function in question; in other words, one is perhaps justified in saying that complexes of action are fixed in the cerebral cortex in relation to the necessity for their performance.

The great majority of people do not follow a special occupation or pursuit which necessitates the frequent fixation of the eyes upon an object above the level of the head. Indeed, one may say that the objects which engage our attention are almost always situated at or below the level of the eyes. When walking we may watch our feet in order to avoid obstacles or glance from side to side to avoid the traffic, but we seldom look upwards for we have no fear of something falling on us or of an attack from above. We rarely look up, we are indeed, not in the habit of doing so, for we do not anticipate the unexpected. An illustration in point is afforded by the children's game of "hunt the thimble," for I am told on good authority that when the object which the children are looking for is placed just above the line of vision, even though in a conspicuous place, it often

escapes detection. So unaccustomed are we to look up for any length of time that when we spend a long morning in a picture gallery we are reminded of our exceptional experience by the aching of the muscles of the neck or by headache. When we look upwards the eyes are commonly "directed into space," and assume a position of parallelism. I take it that the type of headache to which I have just referred is a consequence of "eye strain" determined by the effort called for in producing the degree of sustained convergence, an unaccustomed movement with the eyes in this position, which is required in fixing objects in our near vicinity situated above the level of the line of vision.

But while, when we look upwards, our gaze is almost always so directed for the express purpose of visual fixation, we may look upwards for other reasons. Some of you are probably familiar with the remark addressed by the schoolmaster to the pupil, "You won't find the answer in the ceiling." While the child's action is capable of other possible explanations, you will, I think, agree that some of us look upwards in periods of mental abstraction, presumably in order to give our association of ideas free play by eliminating visual stimuli which might interrupt or interfere with our train of thought. The same result might be as effectively achieved by closure of the eyes, but this is exceptional, possibly because the movement is too obvious, or for other reasons. The possibilities occur to me that the movement of looking upwards in association with mental abstraction may in some instances be determined by a primitive convention, in other words, by the hope of inspiration from above, or that it may be of the nature of a release phenomenon. Both these suggestions appear to me, however, unlikely. My impression is that the movement is voluntary and purposeful; this is certainly so in my own case, although it may become a habit.

The changes in expression associated with mental abstraction on the one hand, and mental concentration on the other, are striking. Among other distinctive features the pupil tends to dilate in states of mental abstraction and to contract during mental concentration. I often recall the words of one of my teachers, Professor John Wyllie, who when examining the eyes with the ophthalmoscope would say to the patient in a musical, dreamy hypnotic tone of voice, "Think of your distant home." There is a children's game which provides another illustration. The child says to his companion, "I can read your thoughts by looking into your eyes." He asks his playmate to think of a number, let us say the number 1, and then to think of another number, say 100. After asking him to repeat this several times he

says to him, "Now think of either one or a hundred and I will tell you the number you are thinking of." In nine cases out of ten he is right, for when one thinks of the larger number one's pupils tend to dilate, while when one thinks of the smaller number they contract. This, however, is by the way. Again, the upward movement, or the direction upwards of the eyes, is a stereotyped characteristic of the suppliant or devotional attitude, as the works of the old masters constantly remind us. The domicile of the Deity is by common consent located in the heavens above, an obvious reason why the eyes should be directed upwards in addressing or approaching the Almighty.

Further, the movement of turning up of the eyes has come to be utilized as a conventional mode of expression. Thus certain individuals are in the habit of turning up their eyes, either in association with or independent of spoken language, in order to express or accentuate their meaning. Thus I overheard a young girl remark to her companions: "Oh, if I had heard her say that I should have turned up my eyes!" What did she mean? My impression is, that she intended thereby to imply disapproval or doubt, for these appeared to me to be the conventional meanings which are attached to and implied by this movement. But I may remind you that, as Darwin [4] points out, different races attach different meanings to the same expressive movement just as they employ different languages with which to express their thoughts. A conventional expressive movement must have an origin. It may be that the upward movement of the eyes, when employed to express disapproval, had its primitive origin in a desire on the part of the individual to avoid seeing that which met with his disapproval, or it was perchance an appeal to the Almighty to help him to resist temptation.

Individuals vary greatly in the extent to which they exhibit or employ facial movements in relation to expression. At the one extreme we find the country yokel with a face so immobile as to suggest the Parkinsonian mask, while the actor, on the other hand, employs expressional movements in an exaggerated form. The similarity between the physiognomy of an illiterate ploughman with an almost complete absence of expressional response, as compared with that of a patient who exhibited a slight degree of encephalitic Parkinsonism, recently afforded me an interesting subject for discussion. In the one case the individual had not acquired the habit of using his muscles of expression, whereas, in the other, the movements of expression were interfered with as a consequence of an organic lesion of the nervous

system. Cerebral activity, beyond doubt, plays an important rôle in the acquisition of expressional movements. One recalls Millet's famous pictures of illiterate peasants working in the fields, carrying out their daily duties with machine-like regularity, with empty minds, and with little opportunity for that intercommunication between man and man which plays such a prominent part in creating and stimulating the thinking brain.

But I would remind you that we are trained from early childhood to control emotional display, and that great differences are met with not only in individuals but also in different races. I recall the words of one of our most famous tragedians to the effect that the great actor passes through three phases—firstly he, so to speak, learns his part; secondly, he comes to feel and live the part; and finally he arrives at a higher sphere when he utilizes all the necessary emotional expressions with telling effect, although these are divorced from any corresponding emotional feeling.

Mimicry and mannerisms.—Expressional movements are used by all of us in varying degree. They constitute up to a point a means of intercommunication, especially in relation to the emotions, and are thus comparable with language. Darwin's references [5] to the case of Laura Bridgman certainly suggest the possibility that in some instances such movements may be innate, but all the probabilities would seem to indicate that they are usually acquired. Custom has attached conventional meanings to certain movements, and these are picked up just as is language—although the child receives no special training unless it be that he is commonly corrected should any tendency to exaggeration be noticed. Whatever be the primary origin of a conventional movement there can, I think, be no question that unconscious mimicry is commonly responsible for its transmission. Some individuals are, however, much more prone to acquire such movements than are others. Striking similarities in expressional behaviour are often noticeable in members of a family and in the case of individuals who are brought much in contact with one another. If one watches a number of persons sitting round a room, listening to speakers who are in turn expressing their views upon a subject in which those present happen to be especially interested, or regarding which they hold strong opinions, one notices on the part of some of the listeners a tendency to adopt a pose very similar to that adopted by the speaker, whereas others, on the contrary, adopt an indifferent or antagonistic attitude. The sympathetic listener would appear to unconsciously assume the posture of the speaker either

in order that he may the better appreciate the mental attitude of the latter, or for the purpose of indicating to the speaker and those present his agreement with the views expressed, or it may be to emphasize to himself or those present his open-mindedness, i.e., his preparedness to give the speaker an impartial hearing—although he may not be prepared to express a positive acquiescence.

When a movement of the kind which is unusual or exaggerated becomes habitual, we term it a *mannerism*. All of you have no doubt been struck by the way in which assistants are apt to acquire the mannerisms of their chiefs. I well remember attending, as a student, a clinical lecture delivered by a distinguished surgeon who had a habit of puffing out his cheeks. His assistant surgeon and clinical tutor stood behind him with folded arms listening to his words, and both were quite unconsciously indulging in the same grimace. This incident provides an illustration of the development of a mannerism by mimicry, the way in which many of the expressional mannerisms are certainly acquired. There are, however, other ways, and the habit of "turning up of the eyes" usually originates, I think, as a purposive act. One may turn up the eyes in order to emphasize one's disapproval, and again, one may acquire the habit of doing so, unconsciously, when one would not express disapproval or doubt in words, either because one does not wish to wound the susceptibilities of the individual, or it may be for other reasons.

When an habitual movement at first purposeful becomes purposeless, it falls into the group of the *tics*. A mannerism becomes a tic when it is dissociated from the conditions or the purpose which originally gave rise to it. We sometimes meet with cases of upward movement of the eyes in which the movement, at first a mannerism, becomes a tic. A case recently met with is of interest in this connection, for it indicates, not only the development of the upward movement of the eyes as a tic, but also raises an unusual point in diagnosis. The patient, a girl, aged 14, had had two major epileptic seizures. Her mother stated that on each occasion the fit had been preceded by turning up of the eyes, but she added that this movement did not strike her as unusual since, she said, the girl often turned up her eyes, especially when preparing her lessons. I suspected that the patient was subject to attacks of *petit mal*, but when I questioned the girl—and I had no reason to doubt her statement—she remarked in words to this effect, "Oh, that's nothing; it is just a habit I have got into. You see, the girls in my class don't always approve of what our school teacher says, and when we disapprove

we just turn up our eyes. There are several girls in the class who do the same thing, and I have just got into the habit of doing it."

The upward movement of the eyes, though not uncommon as a mannerism, is, in my experience, uncommon as a tic, possibly because the movement is so apparent that its occurrence attracts attention, and is thus less likely to be unconsciously imitated.

The eyes commonly deviate to one side during a generalized or unilateral epileptic fit. I cannot say whether they are ever directed upwards or downwards, but I have had no recent opportunity of making observations on the point. It is indeed surprising that a phenomenon may have constantly occurred before one and yet have escaped notice, unless one's attention has been for some special reason directed to it. On the other hand, the relatives and friends of an epileptic patient not unfrequently state that the eyes move upwards in slight attacks, and I have myself seen two patients recently, during attacks of *petit mal*, in which the eyes turned almost directly upwards. More commonly, however, the patient merely stares. I know of no series of observations upon the position of the eyes in epileptic attacks, but I have the impression that the analysis of a collected material directly bearing on this point might prove informative in relation to the seat of origin of the attack, and possibly helpful in prognosis and treatment. The subject is one which, it seems to me, is well worthy of the attention of those who have special facilities for observing patients during attacks.

The cortical representation of the upward movement of the eyes.—The upward, downward and convergent movements of the eyes can all be carried out voluntarily, and must consequently be represented in the cerebral cortex. I know of no reported case, however, in which a fit characterized by upward, downward or convergent movements of the eyes—or a paralysis of these movements—was associated with organic disease of the cerebral hemispheres. Again, those observers who have especially studied the effects of stimulation of the cortex in normal animals and in man have, so far as I know, failed to produce an upward or downward movement of the eyes. Hughlings Jackson [6] advanced an explanation to account for the facts which remains unchallenged. I may remind you that it occurred to Jackson that the lateral movements of the eyes might be so much more powerfully represented in the cortex, than any of the other ocular movements, that they overpower the latter when the eye-area is stimulated. Risien Russell [7], whose experiments were carried out at Jackson's suggestion, stimulated the "eye area" of the cerebral cortex on one side after

previously dividing the internal rectus of the eye on the same side and the external rectus of the eye on the opposite side and found he was then able to elicit upward and downward movements of the eyeballs. He consequently concluded that "Hughlings Jackson's suggestion as to the probable explanation why under ordinary circumstances none but the lateral movements of the eyes to the opposite side can be elicited on excitation of the one cerebral hemisphere, appears to be the correct one. . . The other ocular movements are as truly represented in the cortex as is the lateral movement of the globes to the opposite side; the only difference is a question of degree in their representation, the latter being more powerfully, or more widely, represented than any other ocular movement"

An innate co-ordinated upward movement of the eyes.—My remarks hitherto have been confined to the upward movements of the eyes in so far as they appear to be more or less obviously referable to the cortex of the brain. There are, however, upward movements of the eyeballs, which seem to be more or less directly associated with, or related to, a "centre" situated at a lower level.

According to Preyer, the eyes of the new-born infant do not manifest any apparent co-ordinated movement which cannot be attributed to accident. I have, however, repeatedly noticed a tendency, when an infant was sitting half asleep on its nurse's knee taking a bottle, for the eyeballs to roll upwards exposing the sclerotic, in association with drooping of the lids, just as is the case in the adult who has difficulty in keeping awake. My impression is that this is a definite co-ordinated movement which is present at birth, and Mr. W. P. Kennedy, who has at my suggestion made some observations upon the eyes of infants during the first few days of life, came to the same conclusion. Assuming this observation to be correct, we have here an instance of a co-ordinated movement of the eyes which is no doubt of a lower order and which, unlike the co-ordinated movements determined by vision, is innate. In other words, while the co-ordinated movements of the eyeballs are essential to vision, the co-ordinated upward movement is exceptional in that it is present at birth, and presumably determined by a different type of physiological demand.

An upward movement of the eyes, of which the individual is unaware, occurs under normal conditions when the eyes are firmly closed. Although the phenomenon may not be easy to demonstrate in the healthy person, since it only occurs in forcible contractions of the ocular sphincter, it is very evident in cases of peripheral facial paralysis

or paresis in which the patient cannot close the eye, or in which the upper lid can be passively raised without difficulty. The rolling upwards of the eyes under such circumstances is of some clinical significance in the detection of slight degrees of facial paresis for, as the movement only occurs in association with powerful contractions of the sphincter, its occurrence indicates that the patient is making a real effort to contract the sphincter of the lids, and *vice versâ*. This movement whereby the cornea is brought under the cover of the bony ridge of the eyebrow constitutes, with the associated closure of the eyes and contraction of the pupils, a very beautiful illustration of a protective mechanism.

Blinking.—The act of ordinary blinking or winking is accompanied by a synchronous upward rotation of the eyeball. Thus, according to Ponder and Kennedy [8], "Each blinking movement is associated with a movement of the eyes itself, the point of fixation being moved some 15 degrees, and the eye rapidly jerking upwards and inwards." These observers are of opinion that "the movements (of blinking) are centrally controlled and dependent on intermittent impulses passing from the region of the basal ganglia," and that "the act of blinking is closely related to the 'mental tension' of the subject at the time, the movements constituting in all probability a kind of relief mechanism, whereby nervous energy, otherwise unutilized, passes into a highly facilitated path." In support of this conclusion they remark that "states of abstraction," or more accurately absolute blankness of mind, "and states of intense but internally directed mental activity, such as when a mathematical problem is being solved, are associated with a slow blinking rate." While, on the other hand, "a state of mental tension which has no internal or external outlet, as impotent rage, anxiety necessarily accompanied by inactivity, or excitement which cannot be shown and expressed by physical movements, is accompanied by a rapid blinking rate." What then is the economic purpose of the elevation of the eyeball associated with the act of blinking? The eyelids brush the conjunctival surface like the windscreen wiper of a motor car, and it would seem that the purpose of the associated upward movement of the eyeballs is to aid in the sweeping of the conjunctiva and the disposal of the tears. In this connection it is interesting to note that young infants do not wink. This was my impression from watching my own children during early infancy, but I confess that the significance of the observation had not previously attracted my attention although it may be familiar to some of you. I was consequently much interested to find

that Ponder and Kennedy [9] observed that "True blinks first make their appearance about the age of six months, and even then are infrequent." This observation was, I understand, based on a single case and, if so, cannot be accepted as necessarily applying to all. It is interesting in this connection to compare the time when the infant first sheds tears. According to Preyer [10], two cases were observed by Darwin in which the eyes were wet with tears for the first time at the end of the third and ninth weeks, while in another case tears flowed down the cheeks at the end of the sixth week. Then again, one of Darwin's children first shed tears in crying in the twentieth week, while Champney's child shed tears for the first time in her fourteenth week. These facts would seem to suggest a time developmental relationship between the act of blinking and the shedding of tears. The question appears to me to be of interest in relation to emotional expression. I must, however, leave it at this point.

The position of the eyes during sleep.—The eyes roll upwards during sleep. We can all no doubt recall occasions when we have watched a companion whose eyes, to use a popular expression, were "heavy with sleep." We have seen his eyelids slowly droop, the eyeballs move upwards so that the sclerotic only is visible, the nodding movements of the head and the sudden start as the muscles generally relax, a series of phenomena which continues to recur despite all the efforts of the individual to keep awake. The upward turning of the eyes during sleep is not always easy to demonstrate, for on attempting to raise the upper eyelid of a sleeping person, unless he happens to be in a deep sleep, one may meet with a certain degree of resistance. This may perhaps be explained by a persistance of tone in the sphincter. My impression is, however, that there is an actual reflex contraction of the muscle which probably forms part of the reflex defence mechanism of sleep. Dr. W. R. Russell, my house-physician, who examined ten sleeping patients, found that in all there was some slight resistance on attempting to elevate the upper lid, that in all the pupils were contracted, that in six the eyes were directed straight forwards, in two slightly upwards and in two markedly upwards, although in one of these they almost immediately came down. These observations suggest that the contraction of the pupil occurs in lighter degrees of sleep than does the upward movement of the eyes, and that the sphincter tone or reflex persists, in some cases at all events, even when the pupils are contracted and the eyes turned upwards. Very exceptionally individuals sleep with their eyes open. The phenomenon is certainly very unusual for

Dr. Russell, who recently made inquiries throughout the hospital, was unable to find at the time a single instance. My two ward sisters tell me that, to the best of their recollection, the eyes are always turned upwards under such circumstances. According to Gowers [11], the eye remains partly open in cases of complete peripheral facial paralysis during sleep. I have, however, a case of complete peripheral facial palsy at present under my care, the only case of the kind I have had an opportunity of observing since my attention was directed to the point, in which the eye is completely closed during sleep although the patient cannot voluntarily close it when he is awake.

One sometimes hears the expression "the cadaveric position of the eyes," and I have heard it stated that the eyes turn upwards after death. While the eyes may turn up during the act of dying, there can be no doubt that after death the axes of the eyeballs are almost always if not invariably, in the normal position of rest, i.e., in the horizontal plane.

The position of the eyes during surgical anæsthesia.—A possible comparison between the position of the eyeballs in sleep and in surgical anæsthesia naturally suggests itself. Very little attention appears to have been paid to the eye movements in anæsthesia, probably because, as Dr. John H. Gibbs writes me, "These ocular movements are of very little practical use to the anæsthetist." The three anæsthetists with whom I have discussed the question are all in agreement that in the great majority of cases the globes remain central. According to Dr. Gibbs, "One meets with lateral deviation, but it is much less common than upward, whilst downward movement is, in my experience, very rare." Again he remarks, "With ethyl chloride, and with no other anæsthetic agent, I have repeatedly seen vertical nystagmus which passes off with deeper anæsthesia." Dr. J. S. Ross writes me, "During surgical anæsthesia the eyelids are usually almost closed with the eyeballs at rest and looking forward and slightly downwards. An extreme rotation downwards may usually be taken as a sign of a very deep anæsthesia." Dr. Torrance Thomson tells me that his impression is that a downward deviation of the eyes occurs more commonly than upward or lateral movements. Thus it would appear that deviation of the eyes is exceptional under anæsthesia, and that the behaviour of the eyes differs not only according to the depth of anæsthesia but also according to the anæsthetic which is used.

No series of observations has been published, so far as I am aware, upon the position of the eyeballs in man during anæsthesia. Further,

it is difficult to distinguish between voluntary and reflex movements. Risien Russell [7] has, however, I may remind you, recorded his observations upon the position of the eyes in dogs anæsthetized with chloroform and ether. He makes no reference to upward or downward movements of the eyes, although he noted a lateral deviation with great regularity, in most of the dogs to the right in a few to the left, about the time when surgical anæsthesia was about to be established. Dr. Hughlings Jackson's comments [6] upon these observations are of much interest.

The representation of the upward movement of the eyes in the brain-stem.—An isolated loss of the associated upward movement of the eyes is practically pathognomonic of a lesion within the substance of the brain-stem. There are, as is well known, many recorded observations in support of this conclusion. Evidence is afforded both by cases in which loss of the upward movement was met in association with a demonstrable post-mortem lesion, commonly a tumour, and by cases in which the accompanying symptoms, in the absence of post-mortem evidence, were clearly indicative of a lesion in this situation. Conversely there is, so far as I am aware, no evidence that a lesion in any other part of the nervous system, the brain-stem being intact, ever results in an isolated loss of the associated upward movement of the eyes. Spiller [12] analysed the available facts in 1905. The lesion, in those cases which have been pathologically confirmed, was "in the vicinity of the oculomotor nucleus" and involved the adjacent region about the aqueduct of Sylvius. Although the clinical evidence points to an area in the brain-stem destruction of which results in loss of upward movement of the eyes, we possess, so far as I am aware, no more precise knowledge of the structural mechanism to which this specific disturbance can be definitely relegated. Lethargic encephalitis is in my experience much the most common cause of isolated paralysis of the upward movement.

Spurious paralysis of the upward movement of the eyes.—But while paralysis of the upward movement of the eyes is a distinctive focal sign, I have met with several individuals who had difficulty in raising, or indeed were quite unable to raise, the eyes above the horizontal when asked to do so, although they were able to elevate the eyes to the full extent if asked to follow the finger upwards. The apparent loss of power cannot be explained by inattention, or perhaps I should say by inattention only, since it was confined to the upward movement. Fixation of the eyes above the horizontal with the head erect is an

unaccustomed movement, and the infrequency with which we are called upon to direct our eyes upwards may account for this observation. The disability manifested by these patients would seem to be comparable with the known difficulty of converging without having an object on which to fix the eyes. Again, we find an analogy in the difficulty some individuals have in wrinkling the forehead, and even in expanding the thorax when asked to do so. Duane [13] writes as follows with reference to loss of the upward movement: "It is probable that some of the cases reported are spurious, the power of movement being present but never put forth. This was so in one of my cases, in which repeated examinations extending over several years always showed complete inability of the eyes to go upwards above the horizontal plane, and yet Graefe's test proved that they could really turn up at least 30°, and subsequently the patient regained the power of spontaneous movement."

Obviously it is important from the point of view of diagnosis to distinguish a spurious paralysis in which the patient does not move his eyes upwards when asked to do so, although there is no actual loss of power, from an actual paralysis of the upward movement of the eyes. Accompanying signs such as other extra- or intra-ocular palsies, absence of blinking, or the presence of vertical nystagmus on looking upwards, would point to a true paralysis. If a paralysis of the upward movement ever results from a lesion situated above the level of the brain-stem "centre" one might conceivably expect to find in such a case that the eyes rolled upwards when the eyelids were tightly closed. I have never, however, met with an instance of the kind although I have on several occasions examined cases of paralysis of the upward movement with this possibility in view.

Turning up of the eyes in encephalitic Parkinsonism.—Involuntary turning up of the eyes occurs in some cases of epidemic encephalitis. This symptom has attracted considerable attention during the past two or three years. There would appear to be no reference to it in the literature prior to 1923. According to Wimmer [14] movements of the kind were observed by Bing in only three in a total of about three hundred encephalitic patients, while Wimmer met with five instances in a somewhat similar material. I have notes of eleven cases in which such movements of the eye were remarked on. These figures, no doubt, underestimate the relative prevalence of the phenomenon since the movements in their slighter degrees cause little or no inconvenience. There were associated manifestations of Parkinsonism in all my cases and this would appear to be the universal experience. The symptom is

pathognomonic and is sometimes of diagnostic importance, since the accompanying indications of Parkinsonism may be so slight that the true nature of the malady might otherwise pass unrecognized unless by an experienced observer.

Two of my patients complained of "turning up of the eyes" as the only symptom causing disability or inconvenience. Among my eleven cases the eyes moved more or less directly upwards in eight; in one case the eyes were directed upwards at one period and downwards at another; in one they were sometimes directed upwards and sometimes downwards, while in another the movements were to the right and slightly upwards. No paralysis of upward movement was observed at the time of examination in the three cases in which downward and lateral movements occurred.

The deviation of the eyes may be momentary. On the other hand the eyes may be directed upwards for hours at a time although it may be possible for the patient to bring them down and keep them down for short periods by exercising "an awful effort," to use the expression employed by one of my patients. One patient stated that the eyes tended to turn upwards when he was in the house, whereas when he was out of doors they turned downwards, although they were apt to roll upwards if he sat for some time watching a game of tennis. Another patient remarked that she had noticed a special tendency for the eyes to turn up if she looked at a bright light. This patient told me that she found that the eyes soon resumed their normal position if she lay down.

Associated blepharospasm was observed in two of my cases, and two other patients stated that they were unable to open their eyes during an attack, possibly for the same reason. On the other hand one patient said that he was unable to close the eyes during an attack. In more than one case there were vertical nystagmoid movements. A backward movement of the head, in association with the upward movements of the eyes, was noted in one case, but I have not personally met with any instances, such as have been reported by Wimmer and others, in which there were widespread spasmodic movements of the face, limbs and trunk.

Fatigue and emotion are factors which appear to play a rôle in exciting the attacks. Thus in most of my cases the patient stated that the movements were especially apt to occur towards the latter part of the day or when he or she felt physically tired. Several patients remarked that the movements became exaggerated when attention was

directed to them, or when they felt self-conscious or agitated. Thus one girl found that her worst periods were when she sat down to a meal, while a man complained that the upward movements were most troublesome when he was in a crowd. Again, I was struck by the fact when examining one of my patients, the case in which the deviation was lateral, although this is no doubt mere coincidence, that while she was unable to maintain central fixation when asked to do so the movements of the eyes disappeared when I engaged her in conversation. Further, this patient remarked that the movements did not occur when she was reading or sewing. Hyoscine has sometimes an undoubted effect in ameliorating, and indeed may actually control, the movements.

The nature and manner of production of these movements is a question of much interest. Since they have only been observed, so far as I am aware, in association with the Parkinsonian manifestations of epidemic encephalitis, their dependence upon the same pathological process is unquestionable. It is tempting to suppose that the upward movements of the eyes may be of the nature of a release phenomenon, and that as the lateral movements are preponderatingly represented in the cortex, so in the brain-stem the representation of the vertical movements predominates. On the other hand the direction of the movements is not always upwards or downwards, while diplopia is sometimes complained of during the attacks. Again, a variety of tic-like movements of the face, tongue and jaws, are met with in these cases of encephalitic Parkinsonism. It is difficult to avoid the conclusion that an irritative process plays a prominent part in their production.

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NOTE ON THE PAIN SENSATIONS WHICH ACCOMPANY DEEP PUNCTURES.

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DURING the course of determinations of tissue temperatures [Bazett and McGlone, 1927] some hundreds of deep punctures into the tissues have been made, while in other experiments repeated venepunctures have been performed [Dreyer, Bazett and Pierce, 1920], and one of us has also been a subject for arterial puncture on several occasions. These experiments allow certain generalizations to be made in regard to the sensations experienced, particularly as to the similarity of the peculiar sensations which usually accompany arterial puncture to those which occasionally are induced by venepuncture, and which are commonly observed when a needle exerts any tension on the deep fascia. These sensations have a definite character and may perhaps help to elucidate the sensory symptoms of some neuralgias. They are also of interest in relation to the study of nerve distribution to the vessels made by Woollard [1927].

Arterial puncture.—The sensations which accompany such punctures are well recognized. When the needle reaches the vessel wall, a dull aching sensation is felt which is much less acute than is a simple puncture of the dermis; the pain, however, is much less bearable. It is diffuse, is often referred to some other position (usually one more distal), and the aching sensation is often associated with uncontrollable reflex reactions; the subject may complain of a sudden sensation of warmth, may break out into a sweat, may then feel cold, faint, or actually lose consciousness. The sensations may be accompanied by nausea, and in some individuals by a desire to defæcate. There is a similarity to the sensations experienced with testicular injuries; though the actual pain is less intense, the reflex changes of an autonomic type may be just as pronounced. As is well known, the sensations accompanying

punctures of the brachial artery are less intense than those experienced when the radial artery is pierced; this has been very noticeable in our experiments. We have had no experience with puncture of the femoral artery in the groin. In two experiments, where the puncture of this vessel was attempted with a thermocouple needle, no such sensation was experienced at all, but it remained doubtful whether the needle ever entered either the vessel or its wall.

When the muscles, and less commonly the dermis, are punctured the needle may occasionally enter a small artery; with thermocouple needles such an event may be indicated by the occurrence of bleeding along the tract (outside the needle), even when quite small needles are employed. When a vessel within the dermal area has been punctured during the introduction of a thermocouple needle (tangentially or obliquely directed), an unusual degree of pain commonly accompanies the injury, and there is produced an inflammatory hyperæmia greater than the average in area and intensity. When, however, such a hæmorrhage has occurred after the needle has pierced the deep fascia, and where the blood presumably has escaped from some intramuscular vessel, the pain has been of a dull aching character, not easily bearable, and reflex responses of the fainting type have been in evidence. In fact, if a puncture produced such intense symptoms that a needle had to be withdrawn (though introduced in a subject thoroughly inured to the procedure), indications of puncture of a deep vessel, in the form of hæmorrhage along the needle tract, was rarely absent. It seems, therefore, that puncture of the smaller vessels, with the possible exception of those in the dermis, is accompanied by more severe sensations and symptoms than is puncture of the larger vessels.

Venepunctures.—The sensations experienced are usually those of dermal puncture alone, and the vessel puncture gives no sensation. Occasionally, however, a small nerve appears to be affected (in our experience in 2 or 3 per cent. of the punctures); the sensations experienced are then similar to those of arterial puncture, but much less severe. Such sensations have been least rare when a vein has been punctured near a valve where muscular contractability and the supply of nerves might be anticipated to be maximal.

Deep punctures on the extensor aspect of the fore-arm.

Dermis.—Punctures made with a needle at right angles to the surface are accompanied with a simple pricking sensation of an acute type, which subsides at once and which is accompanied by a slight local inflammatory

reaction, but by little or no hyperæmia except in the immediate neighbourhood. If, however, the needle be introduced tangentially, the sensations are more noticeable, last longer, and an inflammatory hyperæmia may often be distinguished over an area of one or more centimetres diameter. The pain is irritating, but not nauseating. The inflammatory reaction and the pain are perhaps more marked when the needle travels parallel to the surface for some distance near the junction of the dermis and fat, but unquestionably there is a definite relationship in any one subject between the severity of the painful sensation and the degree of the inflammatory hyperæmia.

The subcutaneous fat.—A needle introduced at right angles to the surface can usually be moved back and forth in the subcutaneous fat with little or no sensation of pain; there are no unpleasant reflex responses, nor are inflammatory reactions much in evidence.

The deep fascia.—As soon as the needle reaches the neighbourhood of the deep fascia, the subject notices sensations of the type already described in relation to arterial puncture, though, when the needle is very sharp, the sensations are occasionally absent. The sensations are usually only moderately severe and appear to arise when the needle point is in close proximity to, or touching, the external aspect of the fascia; they subside rapidly once the fascia has been punctured. If, however, the needle position is such that tension is exerted on the deep fascia, or if the needle be jarred so that impulses are transmitted to the fascia, sensations may continue or return. The sensations are commonly so definite that the subject can identify the fascial plane as readily as he who introduces the needle, and a subject may faint when the fascia is touched, even though he has shown no reaction for a quarter or half an hour previously while the needle has been only in the dermis and fat.

Muscles.—Once the point of the needle is within the muscles, sensations are as a rule absent, unless tension is being exerted on the deep fascia. Intermuscular septa may, however, be encountered and can be recognized by the resistance offered to the needle. Sensations similar to those induced from the deep fascia may accompany their puncture, though they are usually much less definite. The needle may also encounter small blood-vessels with the symptoms already described. We believe we have touched or punctured occasionally the interosseous membrane in the fore-arm; this has been accompanied by symptoms and sensations similar to those produced by arterial puncture.

Periosteum.—The needles have often been introduced till they have

met the radius or ulna bones. Contact of the needle point with the periosteum usually produces sensations of a moderate severity, which closely resemble those already described in relation to arterial puncture.

Inflammatory reactions.—The inflammatory responses serve to demonstrate that the reactions are not of mere psychical origin. In most subjects puncture of the fascia is not accompanied by any marked hyperæmic reaction; however, in some subjects a diffuse hyperæmia makes its appearance as soon as the sensations of fascial puncture have been noted, but not before, even though the needle may have been in position in the subcutaneous tissue for a long time. In all subjects the painful sensations experienced when a small blood-vessel is pierced, and when the periosteum is irritated, is almost invariably accompanied by the rapid development of a diffuse hyperæmia; there is a noticeable parallelism between the severity of the sensation and the degree of hyperæmia in any one subject.

Deep puncture of the inner aspect of the thigh.—The sensations and symptoms resemble those observed with puncture of the fore-arm, except that in the one subject used the reactions following thigh puncture are greater, fascial pain is apt to be more intense, and the puncture of arterial branches with the accompanying intensified reactions seems to be more common. Most of the punctures have been made over the sartorius muscle in the middle third of the thigh. In this region several fibrous septa may be met within the subcutaneous tissue; these may give rise to sensations similar to those given by the deep fascia, but as a rule the sensations are not so marked. Puncture of the fascial plane on the deeper aspect of the sartorius muscle usually produces a definite reaction; if the needle be introduced at the edge of the sartorius muscle, where the two planes join, the needle may continue to meet with obstruction for a considerable distance, and the whole of such a puncture may be accompanied by moderately severe sensations of the type described.

When punctures have been made on the inner aspect of the thigh, just above the knee-joint over the vastus internus muscle, hæmorrhage along the needle tract and severe sensations of the type described have been more common than with punctures in a more proximal position.

Deep puncture of the external aspect of the thigh.—This has only been attempted (on the same subject used for the other experiments on the thigh) on three occasions; in every case the needle had to be removed hurriedly, soon after puncture of the fascia lata, owing to the

violence of the fainting symptoms which have accompanied the aching sensation, and the consequent danger to the thermocouple needles in use. The symptoms have been more violent than those accompanying radial puncture, and have necessitated the subject lying down for ten to fifteen minutes after the withdrawal of the needle.

Discussion.

The difference in the type of pain experienced with puncture of deep fascial planes, or of a small artery, from that which accompanies dermal puncture is most marked, and the sensations resemble those of some forms of neuralgia. It has been noticeable how commonly there has been evidence of puncture of a small artery, where the subject has considered that a small nerve has been injured, and it seems probable that the sensations may arise from the nervous plexi which accompany the vessels. It is noteworthy that Woollard [1927] anticipated, from his anatomical studies, that the vessels should be sensitive and that this property should be most marked in smaller arteries and rare in the veins; this is confirmed experimentally. The association of similar sensations with puncture of the deep fascia may conceivably be associated with the presence of a plexus of arterial vessels just superficial to this membrane [Spalteholz, 1893], or to the presence of a nervous network in the same position [Ruffini, 1905].

The more marked sensations of the smaller vessels may be dependent on the greater supply of medullated nerves to these vessels [Woollard, 1927], but, if so, the relief of pain in patients with Raynaud's disease subjected to sympathectomy by Davis and Kanavel [1926] is not readily explained.

The nerve-endings in the vessel walls need not necessarily normally carry sensory impressions, since they may represent nerve-fibres utilized in vascular dilation through axon-reflexes, and hence in direct continuity with other sensory branches.

Conclusion.

The pain experienced when arteries are punctured is described, and comparison is made with the similar pain experienced when the deep fascia is pierced or exposed to tension, or when the periosteum is injured. The occasional pain experienced when a vein is punctured is of the same type. The sensation is that of a dull ache, not necessarily very painful, though difficult to endure and associated with reflex changes of an autonomic type, such as sweating, fainting, or nausea.

The puncture of the fascia on the outside of the thigh produces much more severe symptoms than a similar puncture of the inner side, and both produce more severe reactions than does puncture of the extensor aspect of the fore-arm.

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A CONTRIBUTION TO THE CLINICAL STUDY OF SCHILDER'S ENCEPHALITIS.

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THE morbid state first fully described by Schilder [6] as encephalitis periaxialis diffusa is now established as a disease, or perhaps as a type of disease, whose features may be recognized with certainty by post-mortem examination. We have thus reached that stage of investigation at which we can take the morbid anatomy as our criterion, and upon the foundation which it provides we may begin to accumulate and sift the clinical and ætiological data.

The basis of the present paper is provided by a case proved by post-mortem examination to have died of Schilder's disease. Its clinical interest depends upon two unusual features. The first and most important is that a sister and brother who are still alive are both suffering from an organic cerebral affection which cannot be classified under any of the orthodox disease headings, whilst of the remaining brothers and sisters, out of a total of ten, five have died at various ages from 7 months to 13 years of supposed meningitis or convulsions. Thus it would appear for the first time evidence is produced which suggests that Schilder's encephalitis may have a familial incidence.

The second point of interest is that the fatal case already referred to was correctly diagnosed during life upon clinical features which have not previously been recorded as characteristic of the disease.

Case 1.—Tommy P. was admitted to Guy's Hospital, under the care of Dr. Cameron, on October 8, 1926, aged 6, with the complaint of difficulty in walking.

It was stated that he did not walk until he was 3 years old, but from then until the age of 6 appeared normal, and went to school at the age of 4. After the death of his mother in May, 1926, it was noticed that he would sometimes sigh and make noises in his sleep, but this was put down to fretting. He continued at school until June, when he was admitted to the Rotherhithe Infirmary on account of general weakness. He was discharged after a fortnight, but readmitted from August 18 to 31. A report states that, during this period,

for the first few days, he was mildly feverish (99° to 100° F.) and was rather listless. It was noted also that at times he showed a curious sighing type of respiration. Subsequently he became steadily weaker, and by the beginning of October was unable to walk or stand.

On admission to Guy's Hospital he had a rather vacant look. Intellectually, he appeared slow. There was no aphasia. The optic discs were normal. The visual fields were full to rough tests. The pupils were equal, regular and central, and reacted normally to light and in accommodation. Ocular movements were full, but nystagmoid jerkings were seen in lateral deviation to right and left. The functions of both fifth nerves were intact. There appeared to be weakness of both sides of the face, but more marked on the left; the palate was raised to the right, and the tongue protruded to the left. The limbs showed general spasticity and weakness, especially on the left side. The deep reflexes were increased, those of the left side being the brisker. The abdominals were absent; the plantar responses extensor on both sides. There was incontinence of urine and fæces. No sensory loss could be detected. There were no abnormalities discovered in any of the other systems.

It was observed that the child had a curious smile, which developed slowly and took an equally long time to fade. The breathing was shallow and interspersed with many sharp sighs.

The rectal temperature was 99° F., pulse 110, and respiration 30.

Further investigations showed a negative Wassermann in blood and spinal fluid. The latter was clear and not under obvious pressure and showed protein 0·025 per cent., cells 1 per c.mm., slight reduction of Fehling's solution, chlorides 0·69 per cent.

No tubercle bacilli could be detected in the fluid, and cultivations remained sterile after seventy-two hours' incubation at 37° C.

The clinical picture presented, therefore, was that of a progressive double hemiplegia in a child of 6. There was no evidence of increased intracranial pressure. The advanced bilateral pyramidal signs without involvement of brain-stem nuclei suggested symmetrical disease in both hemispheres rather than a single central lesion.

Upon this basis the diagnosis of Schilder's encephalitis was therefore made and subsequently adhered to.

In view of the known tendency of this disease to involve the optic radiations, the patient's visual acuity and visual fields were subsequently examined from day to day, as well as his failing mental powers would allow. Unfortunately, no records of these tests were kept, but, in the opinion of the house physician, Mr. H. Trafford, the visual power and range remained normal up till death. The optic discs were also examined from time to time, and were observed by the writer to remain normal, and were so when the child was last seen on the day before death, which occurred on October 21.

Post-mortem examination.—The brain and spinal cord, which showed no external abnormality, were separately removed and placed in formol saline. One or two vertical cuts were made through each cerebral hemisphere so as to allow rapid and complete fixation. After hardening, the brain-stem and cerebellum were detached, and the hemispheres cut horizontally into slabs of about

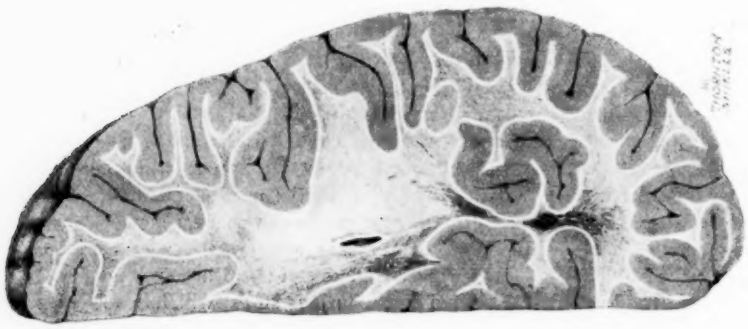

FIG. 1.—Horizontal section through the right cerebral hemisphere just above the lateral ventricle. Brain of Tommy P. Formalin fixed. Unstained.

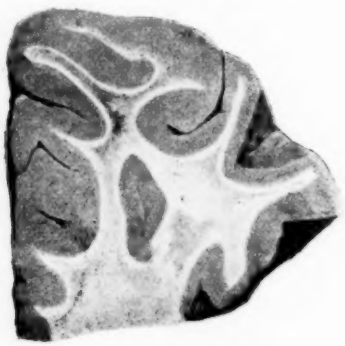

FIG. 2.—Horizontal section of right frontal lobe a few centimetres below the vertex. Brain of Tommy P. Formalin fixed. Unstained.

2 cm. in thickness. The brain-stem, cerebellum and spinal cord were accidentally destroyed before they could be examined.

Macroscopic examination of the hardened slabs at once confirmed the diagnosis of Schilder's encephalitis.

Throughout both hemispheres the white matter showed a diffuse gelatinous degeneration, most advanced in the deeper parts—where in some areas cavities had formed. Where it extended towards the cortex, the degeneration left the subcortical white matter intact, so that this layer stood out as a pale strip

between the grey degenerated area internally and the grey matter of the cortex externally (fig. 1).

The distribution of the areas of degeneration was studied by naked-eye

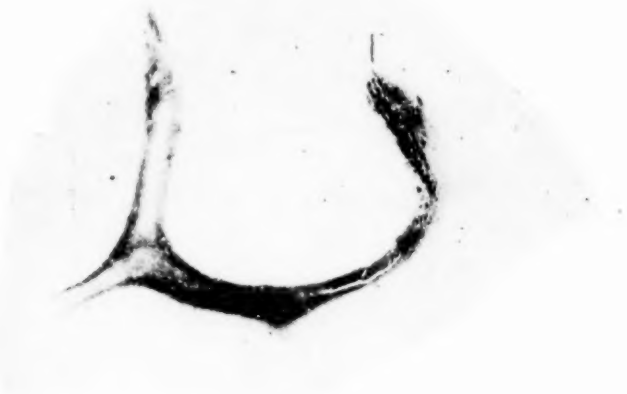

FIG. 3.—Section of the right frontal lobe. Brain of Tommy P. Stained by the Weigert-Pal method. In one part of the section even the subcortical fibres show evidence of demyelination.

FIG. 4. — Section of the left parietal lobe. Brain of Tommy P. Stained by the Weigert-Pal method. The base of the gyrus is severely involved. The demyelination extends throughout its core, leaving a relatively thick surround of well-myelinated fibres subjacent to the cortex.

examination of the serial slabs. The white matter of both frontal lobes was involved throughout their whole extent from vertex to base (fig. 2). The degeneration appeared to be of oldest standing around the anterior cornua of the ventricles, where in parts there was actual loss of substance.

The temporal, parietal and occipital lobes were relatively less affected. The white matter surrounding the ventricles was severely involved, but there were comparatively well-myelinated areas in the centrum ovale, and the white matter of the gyri in those parts did not show the same uniform and clear-cut distinction between a central core of grey degeneration and subcortical strip of normal fibres which was evident throughout the frontal lobes.

Around the posterior cornua of both lateral ventricles, however, the degeneration was more striking than in any other part of the brain, so that in the shrunken condition of the fixed specimen the neighbouring masses of grey matter formed the lips of irregular cavities from which all semblance of white matter had disappeared, except the narrow strip of subcortical fibres.

The disease was, on the whole, symmetrically distributed throughout the two hemispheres.

Microscopic examination confirmed the naked-eye impression of symmetrical demyelination, furthest advanced in degree and extent throughout the frontal lobes. Here in some areas it was difficult to obtain any positive result from the Weigert-Pal method, even the subcortical fibres taking up the stain faintly (fig. 3).

Further back (e.g. in the parietal lobes), the disease process was less advanced. The base or pedicle of the gyrus was severely involved, and the demyelination extended throughout its core, leaving however a relatively thick surround of well-myelinated fibres subjacent to the cortex (fig. 4).

Under the higher powers it was clear that there was swelling and fragmentation of fibres adjacent to the areas of complete degeneration which appeared well stained to the naked eye, and it was impossible to draw any sharp line of demarcation between the healthy and the diseased tissues.

Sections stained with Scharlach R. showed a comparable state. In the zone nearest the cortex the myelin was swollen and fragmented, and there were occasional mononuclear cells with scarlet inclusions in the perivascular spaces. Within this again there was an area in which the remnants of the myelin were represented by yellow-stained granules and droplets lying within a reticulum of swollen mononuclear cells, whilst at the base of the gyrus there were clusters of cells packed with scarlet granules.

Corresponding sections stained by Anderson's modification of the Victoria blue method for neuroglia showed in the subcortical layer an increase of glia fibrils which became thicker and more numerous towards the centre of the gyrus. In the central core was a dense felt-work of fibres running in all directions with many spider cells. Other sections stained by Busch's modification of the Marchi method provided the counterpart to the Weigert-Pal sections, showing the finger-like extension of myelin decay extending from the centrum ovale into the core of the gyrus, towards, but not reaching, the cortex.

The family history.—The father of Tommy P., aged 48, is at work in London. He has enjoyed good health, with the exception of an anthrax pustule some years ago, from which he made a complete recovery. He has had defective vision since the age of 12. It has not been possible to examine him.

The mother, who had previously had no illness of importance, died of pulmonary tuberculosis one year ago.

There have been thirteen children of this marriage, of whom two—besides Tommy—have been examined by the writer.

Concerning the remainder, the data here recorded were obtained in a personal interview with the father.

(1) The eldest child, a male, died at the age of 2 weeks, the cause of death being unknown.

(2) *Case* 2.—Nellie P., the second child, aged 20, is an inmate of Leavesden Mental Hospital, where the writer was enabled to examine her on December 18, 1926, by the kindness of Dr. R. M. Stewart, who has placed the available records of her past history at his disposal.

Her father states from memory that she never walked and was backward and invalid from birth. It was, however, recorded on her case sheet when she was admitted to the Fountain Hospital in March, 1918, at the age of 11, that she had walked at the age of 10 months and talked early. This history was presumably obtained from her mother, now deceased, and is more reliable.

On the same case sheet it is recorded that she was observed at the age of 1½ years and was backward then. From the same record it appears that on admission (aged 11) she was quite helpless, could not stand or sit alone, did not know her age or address, only knew two colours, blue and red, and made no attempt to describe a picture even with help. The note on the physical condition is: "Fair condition, somewhat thin, lower limbs crippled, hand and fore-arm apparently contracted." The circumference of the skull was 20 in.

From this time forward she was under institutional care as an imbecile. Occasional notes of her physical condition record that in February, 1919, "she talks to other children and is remarkably observant. No grip to her fingers." In April, 1921, "on account of all her limbs being crippled she makes no headway, talks readily and is fairly intelligent." In April, 1922, when she was transferred to Leavesden Hospital, the report is: "She is an imbecile without epilepsy. Cannot read or write; can only count up to ten; can recognize simple objects. She is in poor general health; feeble circulation in extremities. Cerebral diplegia: extensor paralysis of fore-arms; spastic paralysis of lower limbs." In February, 1925, the first note is made of her vision: "She is blind and quite helpless, but shows a fair amount of intelligence. Her answers to questions are childish and often irrelevant."

There is no record of her ever having had any epileptic attacks. Her father thinks that her eyesight was normal when she left home in 1918, and that she has only become blind in recent years.

The following notes were made by the writer on December 18, 1926:—

Nellie P., aged 20, admitted Fountain Asylum, 1918. Examined with Dr. R. M. Stewart, December 18, 1926.

From nurses.—She will converse in a childish way with them and with other patients. Takes food slowly out of a spoon, has no dysphagia. Knows

where she is. Recognizes people by their voices. Can see a little. Has not changed much in last year or so, except that she has become more faulty in her habits. No epilepsy.

Examination.—On hearing strange voices became agitated and distressed, crying and shrieking for Nurse Mabel, and saying, "Go away." Articulation clear; voice strong. Can distinguish light from darkness. Told by nurse that the visitors were gone, she said "Go away" when I flashed an electric torch in her face. Optic discs pale and clear cut, the appearance of primary optic atrophy. Pupils dilated and do not react to light. Divergent strabismus, but no definite ocular paresis detected. No nystagmus. Right face weaker than left as she cries. Sensation not examined for want of co-operation.

Motor system: As she lies in bed the head is somewhat retracted, but this is less evident when she is sat up, and it is difficult to distinguish between resistance and true rigidity. The upper limbs are symmetrically fixed in the following position: Adduction at the shoulders. Extreme flexion at the elbows. Extreme pronation at the wrists. Extreme flexion at the wrists and metacarpo-phalangeal joints. Extension of inter-phalangeal joints. Adduction of thumbs. The rigidity appears to be of the clasp-knife type, but voluntary resistance and contracture make examination difficult. There is apparently no voluntary power, but handling the limbs produces flexor spasms. No wasting.

Trunk: Nothing abnormal noted.

Lower limbs: Both semiflexed at hips, and knees and ankles with toes flexed and curled downwards.

Reflexes: Tapping upper limbs provoked some clonic spasms of biceps. Abdominals not elicited—but interfered with by crying. In the lower limbs tapping produces clonic spasm of calf muscles; no other reflexes obtained.

Sphincters: Wet and dirty.

General physique.—Head and trunk normally developed. Pubic hair normal. Limbs small as in an infantile hemiplegia, with cyanosed extremities. Extreme pyorrhœa. Liver and spleen not felt. Kyphosis of lumbar spine. Chest not examined on account of struggling.

(3) The third child, Freddie P., died recently at the age of 18, of pulmonary tuberculosis.

(4) The fourth child, a girl, is said to have died of meningitis at the age of 13 months. No clinical data available. No autopsy.

(5) Lily P. is said to have died of meningitis, aged 13. No clinical data. No autopsy.

(6) Jessie P., aged 15, is alive and well and keeps house for her father.

(7) *Case* 3.—William P., aged 11, was first admitted to Guy's Hospital at the age of 3, when notes were made of his condition, and has been under the observation of the writer for the past five years.

As an infant he developed normally, but he began to suffer from fits at the age of 2, this being attributed at the time to a slight head injury. Subsequently there were convulsive attacks at irregular intervals. According to the mother, these would usually begin with twitching of the right face, arm

and leg; the convulsions would then become generalized and the patient would lose consciousness.

He was first admitted to Guy's Hospital in April, 1919, during one of his attacks, which had begun as usual with right-sided convulsive movements. On admission he was unconscious, but quickly recovered.

Under observation he was thought to be mentally backward and nervous. No localizing signs were found, and he was discharged with the diagnosis of Jacksonian epilepsy.

He was readmitted in July, 1919, and was said on this occasion to have had a left-sided Jacksonian attack. On this occasion he was seen by Mr. H. L. Eason, who noted defective vision, especially in the lower fields, and optic atrophy. The Wassermann was negative in the blood. The diagnosis of cerebral tumour, unlocalized, was made, and he was discharged.

Following this, he had no more attacks until September 14, 1922. On that day there was a series of convulsions, each beginning with twitching of the right angle of his mouth, spreading over the right side of the face, subsequently affecting the right arm and leg. He was admitted unconscious during one of these attacks. On the next day (September 15, 1922) the fits had ceased, but he was still unconscious. The deep reflexes on the right side were increased and the right plantar response extensor. On the 16th he was conscious, but remembered nothing since going to bed on the 14th. The reflexes were as before.

The writer saw him for the first time on September 22, 1922, and made the following notes:—

Cerebration: Odd, over-active. Speech: normal.

Special senses: Vision: acuity very fair to rough tests. Fields: some peripheral construction, especially on right. Discs white and clear-edged, some pigment disturbance around, vessels a fair size, cups not very distinct. I think a primary atrophy.

Cranial Nerves: Pupils dilated with atropine, neither eye turns in very well on conjugate deviation. Slight weakness of right face on emotional movement, otherwise normal.

Sensation: No defect of sensation.

Motor System: Left grip better than right, but says he is left-handed.

Reflexes: All tendon-jerks present and equal on the two sides. Abdominals upper and lower present and equal on the two sides. Plantar responses: right doubtful, left flexor.

Sphincters: No disturbance of sphincter control.

Gait: Walks on a rather wide base and is unsteady.

Impression: Cerebro-cerebellar atrophy.

He was again admitted in February, 1925, and was then reported to have had mild fits once a fortnight, or oftener, since his discharge from hospital in 1922. The account given by his mother was that on these occasions, if playing in the street, he would run inside and lie down; after a minute or two, twitching would begin in the right side of the face and extend to the arm and leg; this would continue for a few minutes; after that he would feel quite well

again and ready to go out to play. Consciousness was not always lost in these attacks. On the day of admission an attack of this kind was followed by a series of fits, during one of which he was admitted. Chloroform was given to control the convulsions, which subsided, leaving him dazed and confused. He was discharged ten days later.

Following this, the fits continued at irregular intervals, but lost their Jacksonian character and became generalized. The story now is, that he is found unconscious, having fallen down.

He was readmitted in December, 1926, for a month's observation, during which he had one brief attack; he was heard to grunt but, by the time the nurse reached his bed, there was no convulsive movement and he appeared dazed, but was not unconscious.

On examination the physical signs were as before. Physically and mentally he is underdeveloped, and the optic discs show primary atrophy with general constriction of the fields. An occasional extensor response can be obtained on both sides; otherwise there are no abnormal physical signs.

The Wassermann is negative in blood and spinal fluid. The spinal fluid contains no excess of protein or cells. X-ray pictures of the skull show no abnormality.

(8) The eighth child, sex unknown, is said to have died of meningitis, aged 7 months. No clinical data or autopsy.

(9) The ninth child, sex unknown, is said to have died of convulsions, aged 9 months. No clinical data or autopsy.

(10) Tommy P. is the case of Schilder's encephalitis recorded above.

(11) A boy, said to have died at nine months of meningitis.

(12) Alfred, aged 3, is said to be alive and well.

(13) Irene, aged 18 months, is said to be alive and well.

Discussion.

The clinical diagnosis in the case of Tommy P. rested mainly upon the exclusion of other possibilities.

The indubitable signs of bilateral pyramidal involvement without any nuclear palsies or other evidence of brain-stem lesion indicated disease of both hemispheres. The absence of any headache, papillœdema, vomiting or hydrocephalus was against the diagnosis of tumour. The normal cerebro-spinal fluid excluded meningitis. These points having been made, the gradually progressive course of the illness suggested a subacute encephalitis. Encephalitis lethargica had then to be considered, but the steady progressive evolution of the disease and the clinical picture of pyramidal involvement were against this. Of known cerebral diseases, therefore, Schilder's encephalitis provided the only satisfactory diagnosis.

It was noteworthy that the visual fields appeared intact, but account

was taken of the cases in which the disease was recorded as having commenced in the frontal lobes (Jakob [3], Walter [8], Kaltenbach [4]).

Comparison of the brain of this case, both in its macroscopic and microscopic appearances, with the descriptions of other writers—especially those of Collier and Greenfield—leaves no room for doubt in the writer's mind as to the pathological diagnosis.

The question next arises whether the other two members of this family who are suffering from obscure encephalopathies may be included clinically within the bounds of Schilder's disease, as already described by other writers.

In the case of William P. the symptoms are mental retardation, epileptic attacks and optic atrophy. All these symptoms have been described by various authors in proved cases of Schilder's encephalitis.

An additional feature of this case, however, is the apparent arrest of the disease; for during the past five years, while he has been under the writer's close observation, there has been no evidence of its progress, and from the history it is probable that his condition had been stationary for three years before this.

This, however, is no new conception in the story of Schilder's disease. The case reported by P. Marie and Foix in 1914 [5] was that of a girl who was taken ill at the age of 18 with a rapidly progressive quadriplegia reaching its height on the fifteenth day of the illness. Subsequently there was some improvement and she remained in a stationary condition for ten years. She then died—not of an exacerbation of the original disease, as erroneously quoted by Collier and Greenfield [1]—but of pulmonary tuberculosis.

The pathological appearances were characteristic of the disease now known as Schilder's encephalitis, save that the microscopic detail was that of an old-standing lesion.

In a recent paper, Foix and J. Marie [2] again draw attention to this case, and record two others comparable in the points of clinical arrest and pathological scar. One was a boy triplegic, blind, epileptic and imbecile, who died at the age of 14, the brain showing the typical central, symmetrical, grey degeneration with sparing of the subcortical fibres. Clinically his condition had apparently remained stationary since the age of $4\frac{1}{2}$.

The other case, even more striking, is that of a man, aged 65, who was said to have had convulsions at the age of a few months, leaving as their sequel a spastic right lower limb with equinovarus and extensor

plantar response. Post mortem a patch of sclerosis was found in the left centrum ovale; its relations with the paracentral lobule explained the clinical picture. The lesion showed the typical extension into the gyri with sparing of the subcortical fibres.

P. Marie and Foix [5] named the condition which they originally described centrolobar sclerosis. Foix and J. Marie [2], adhering to this term, consider that it represents a pathological picture distinct from that described by Schilder only in so far as centrolobar sclerosis may be regarded as a late or healed stage of encephalitis periaxialis diffusa.

The conception of the disease held by Foix and J. Marie [2] is that it is subacute in its onset and progress, but not necessarily fatal. It may thus leave its victim crippled with a healed scar, or scars, in the white matter of the hemispheres.

This conception opens up new fields for exploration. The whole group of spastic diplegics or hemiplegics, whose symptoms date from a history of convulsions or other nervous illness in infancy but are not progressive, deserves to be re-investigated on the pathological side. The morbid anatomy of these conditions has never been satisfactorily classified, and it is possible that the appearances now recognized as typical of Schilder's disease might not have been apparent to the unaccustomed eye.

The story of Nellie P. raises further possibilities. Her present state of spastic quadriplegia, imbecility and optic atrophy, suits well with the diagnosis of Schilder's encephalitis. The occurrence of optic atrophy, now several times reported in proved cases of the disease, must depend, as Stewart, Greenfield and Blandy [7] point out, upon its distribution, and is therefore as much an accident of localization as the occipital blindness with normal pupillary reactions which has been such a striking feature of other cases.

Taking the history on the case record to be correct, and assuming that Nellie walked at 10 months and talked early, the clinical symptoms of the disease must have appeared at the end of her first year and have progressed fairly rapidly, in that she was observed at the age of 18 months and was backward then.

It seems, however, both from the case records and the observations of the parents, that the disease has been gradually progressive throughout the whole period of nine years during which she has been under institutional care. For in 1918 there is mention only of hand and fore-arm contracted; in 1919 there is no grip to her fingers; in 1921 all her limbs are crippled but she is fairly intelligent. In 1922 she can

recognize simple objects (presumably visual recognition). In 1925 she is blind. Her father is certain in his own mind that the loss of vision has been a development only of recent years.

If, then, this case is to be regarded as an example of Schilder's encephalitis, we shall have to add a chronic progressive type of the disease to our list of clinical varieties.

Summary.

A case of Schilder's encephalitis is described in which the diagnosis was made during life and proved post mortem.

The family history of this case shows that, of the twelve brothers and sisters, one died in infancy from unknown causes; one died in adult life from pulmonary tuberculosis; three are alive and well; five died in infancy or childhood from causes notified as "meningitis" or "convulsions" without further evidence of the clinical or pathological state. The other two, a girl aged 20 and a boy aged 11, present symptoms of cerebral disease which are described. These symptoms are, it is suggested, compatible with the clinical diagnosis of Schilder's encephalitis.

Evidence is thus produced which suggests that Schilder's encephalitis may have a familial incidence.

I have to thank Dr. Hector Cameron for permission to record the case admitted under his care, and Dr. J. Godwin Greenfield for his invaluable assistance and advice.

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A CONTRIBUTION TO THE PATHOLOGY OF HEMICHOREA.[1]

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MOST of the authors dealing with the problem of the choreic syndrome have adopted Wollenberg's ætiological classification of its different clinical manifestations. A more recent minor modification is that of Lotmar [4], who divided them into three groups :—

(1) The chronic progressive form, hereditary, with progressive dementia = Huntington's chorea.

(2) The infectious and toxic form.

(3) Macroscopic, localized lesions which may involve the cerebellum, the anterior cerebellar peduncles, the red nucleus, the body of Luys, the thalamus or the striate body.

C. and O. Vogt [6], [7], tried to correlate the clinical syndromes with pathological entities. According to their classification of extrapyramidal diseases, the following structural changes will produce chorea :—

(1) Progressive status fibrosus of the striate body.

A. Without severe degeneration of the pallidum and the correlated mid-brain nuclei. Clinically : Progressive bilateral chorea.

(*a*) If this process is localized in the striate body it will reveal itself clinically as isolated chorea.

(*b*) If connected with a diffuse necrosis of ganglion cells in the rest of the brain, especially those of lamina IV of the cortex, the clinical picture of Huntington's chorea is produced.

(*c*) As subsyndrome of general paralysis : Chorea combined with other symptoms of general paralysis.

B. Combined with severe disease of the pallidum : Chorea with subsequent rigidity.

[1] Presented at the Annual Meeting of American Neuropathologists' Club, Atlantic City, N.J., May, 1927. From the Neuropathology Laboratory, Montefiore Hospital, New York.

(2) Status subinflammatorius (encephalitis).

A. Encephalitis of unknown origin in striatum. Clinically: The progressive striate syndrome.

B. Encephalitis epidemica. Clinically: The striate and pallidum syndrome.

C. Sydenham's chorea with a predilection for the striate body. Clinically: Chorea.

(3) Macroscopic, localized vascular disease in the striate body of adult people. Clinically: Hemichorea.

Cases of the last group should help us in solving the problem of the localization of the different forms of extrapyramidal disturbances of motility. As has been mentioned above, such macroscopic localized lesions may involve any part of the cerebellum-nucleus ruber-thalamus-striatum-pallidum pathway, and produce the clinical picture of hemichorea. C. and O. Vogt presented among their material only one case with vascular lesion of the striate body associated with hemichorea. Case 34 (Liepmann's case, Elizabeth L.) :—

An unmarried woman, aged 42, was transferred to an insane asylum because of hallucinatory dementia. At 67 she suffered from an attack of hemichorea which first involved the right face and arm, and five months later also the right leg. The tendon reflexes were very lively at this time. Two years later, two weeks before death, the patient had a left-sided hemiplegia. From the case report it may be concluded that there was a slight degree of rigidity. Marked arteriosclerosis of the cerebral vessels was found. The head of the caudate nucleus and the oral part of the putamen were replaced by an old cyst, which also had partly destroyed the anterior limb of the internal capsule.

A second case is briefly mentioned by Jakob [2] :—

Sudden onset of choreic movements in the right face in a case of psychosis of arteriosclerotic origin. Anatomically, the cystic remains of softening in the ventral and oral part of the head of the left caudate nucleus were found.

A third case of vascular lesion of the striate body with hemichorea was reported by Lloyd and Winkelmann [3] :—

A patient with a right hemiplegia of three years' duration developed a left hemichorea three weeks before death. Anatomically: Generalized arteriosclerosis of the brain was reported. An old focus of thrombotic softening was found in the left internal capsule, and another focus of more recent origin in the right hemisphere which involved a narrow strip along the inner margin of the putamen, but extended into the outer portion of the globus pallidus. The internal capsule was involved to a very small degree in its

posterior portion by a hæmorrhage, which impinged upon the corpus Luysi and extended into the pes pedunculi. Both striate bodies were atrophic and showed poverty in medullated fibres, especially on the right side. Both thalami contained multiple areas of softening and hæmorrhage. The nucleus ruber and substantia nigra were normal.

A fourth and a fifth case belonging to this group are found in the French literature. Case 4, reported by Austregesilo and Galotti [1], was that of a left hemiparesis with right hemichorea. Anatomically: An irregular focus of softening was present in the head of the left caudate nucleus, with complete destruction of the nervous tissue and new formation of many capillaries. The internal capsule and the rest of the brain were intact.

Case 5 is that of Souques and Bertrand [5]:—

A woman, who died at the age of 54, had suffered at 4 years of age from an infectious disease, which was followed by a left hemiplegia and a left hemichorea. Contractures and a positive Babinski developed. Anatomically: A degeneration of part of the right putamen and the caudate nucleus was found. The anterior part of the corpus callosum was destroyed. The corona radiata showed paling of its upper part in myelin-stained sections, and destruction of fibres in its deeper layers. The anterior third of the posterior limb of the internal capsule was involved, and the anterior peduncle of the right thalamus was also cut by an area of degeneration. The pallidum I was affected in its external and posterior third. The body of Luys was atrophic and almost unrecognizable.

The rarity of localized macroscopic vascular diseases in the striate body with hemichorea may justify the report of the following case:—

M. L., aged 58, a Russian Jew, married at the age of 35, and had seven children, all living and well. In 1911, at the age of 43, patient was suffering from diabetes mellitus, which never attained a severe degree during the following eight years. In 1919, amputation of the second toe of the left foot and of the first phalanx of the second toe of the right foot was performed. In June, 1926, patient complained of general weakness and severe pain in the toes. On August 13, 1926, he was admitted to the Post-graduate Hospital, where, on August 20, slight twitching movements of the right face were noticed. These movements rapidly spread to the right arm and shoulder, and later increased in intensity and involved the right leg. On September 20 they had attained such an intensity as to become alarming. On October 5, 1926, the patient was transferred to Montefiore Hospital.

Neurological examination.—Constant twitching and grimacing of the right side of the face. Eye muscles normal. Pupils irregular, right larger than left; sluggish reaction to light, but normal contraction on accommodation. The left corneal and conjunctival reflexes are sluggish. There is a slight drooping of the left angle of the mouth. The uvula deviates to the left; the pharyngeal reflex

is absent. The tongue deviates to the left, but is not tremulous. The right arm is held in adduction, the elbow in flexion, and the hand in partial flexion with fingers contracted. The frequent movements of the arm are jerky in character and choreiform, while the movements of the right hand and fingers, like those of the facial muscles, are often slower and worm-like. There is atrophy of the intrinsic muscles of both hands, more marked on the right. The muscular power of the right arm is diminished; the patient is unable to raise this limb. There is no fixation in the right leg; wasting is present in the

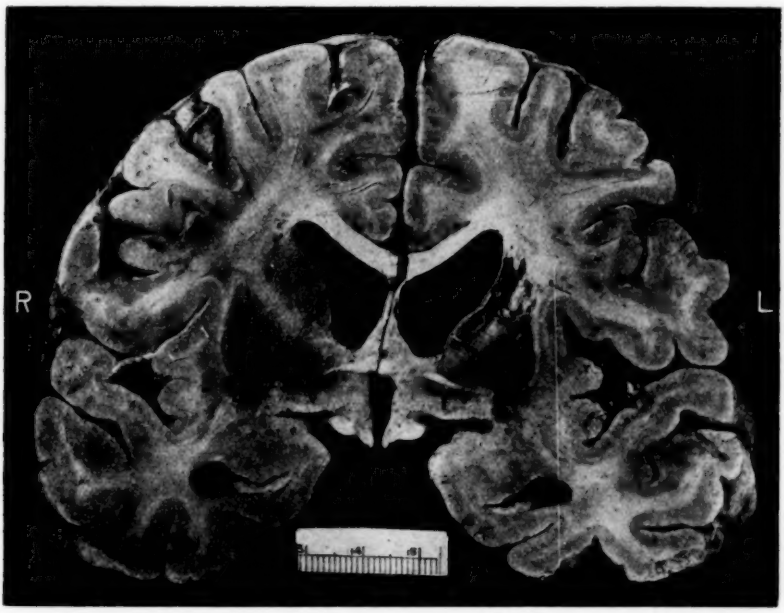

FIG. 1.—Vertical section through brain at the level of the anterior commissure. Cyst formation with destruction of part of caudate nucleus, putamen and internal capsule on left side.

muscles in both legs, more pronounced in the feet, but muscular power is not diminished and both legs can be raised. The movements of the right leg are jerky. The tendon reflexes are absent on the right side, present on the left. There are no signs of involvement of the pyramidal tract. The abdominal reflexes could not be obtained on admission, but on October 25 they were present on both sides, but more lively on the right. Proprioceptive sensations were absent in both lower extremities. Vibratory sense was greatly diminished below the eleventh dorsal root area, and there was hypæsthesia and hypalgesia below the first lumbar area. The mental state was normal. Spinal fluid

FIG. 2.—Vertical section through brain. Loyez stain. Atrophy and demyelination of left striate body and pallidum.

FIG. 3.—Microscopical vertical section through brain, approximately 15 mm. frontal of figure 1. Loyez stain. Cyst formation with partly destroyed striate body and internal capsule

contained 20 cells, a trace of globulin and sugar 4 plus. Blood-sugar was 170 mg., and the blood-pressure 200/90.

Course of the disease.—On November 3 a gangrene of the right second toe

FIG. 4.—Microscopical section through brain, approximately 3 mm. occipital from mammillary bodies. Loyez stain. Atrophy and demyelination of left striate body and pallidum.

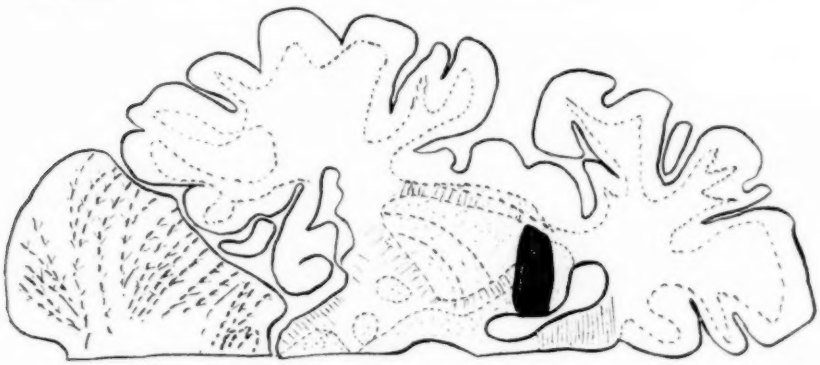

FIG. 5.—Diagram of horizontal section through brain to show approximate extension of cyst in anterior-posterior direction.

developed, first dry, then moist and spreading. On January 1, 1927, amputation of the right lower leg had to be performed. Patient died on January 12, 1927, from pneumonia.

Anatomical report.—Leptomeningitis at the convexities of the brain, with

arteriosclerosis of medium degree of the vessels at the base. The left side of the cerebellum is smaller than the right; the vessels covering the left cerebellar hemisphere are not as prominent as those on the right side. The brain looks smaller than normal, but weighs 1,100 grm. The convolutions throughout are slightly atrophic. The lateral ventricles are enlarged, the left more than the right. There is a cyst formation with destruction of nearly the whole anterior third of the left caudate nucleus, approximately the upper third of the anterior limb of the internal capsule and the upper part of the oral pole of the putamen (fig. 1) The remaining parts of the left striatum and left pallidum are reduced in size. The corresponding basal ganglia and the thalamus on the right side appear normal. In sections through the whole brain stained by Loyez' method, the atrophy of the left striatum and pallidum is still more evident (figs. 2, 3, 4). Drawings were made of these nuclei at a linear enlargement of 1 : 9, and their planes were measured in corresponding sections of both sides with the planimeter. The following table gives the averages of twelve measurements, corresponding to sections taken from the neighbourhood of the section shown in fig. 4. The numbers indicate square inches at a linear enlargement of 1 : 9 :—

TABLE.

	Plane in square inches		Planimetric index		Left nucleus in per cent. of right
	Right	Left	Right	Left	
Caudate nucleus ..	5·760	3·484 ..	26·3	26·9 ..	60·6 per cent.
Putamen	11·254	6·216 ..	51·5	48·0 ..	55·5 ,,
Pallidum I	3·328	2·272 ..	15·2	17·5 ..	68·0 ,,
Pallidum II	1·528	0·982 ..	7·0	7·6 ..	64·4 ,,
Total ..	21·870	12·954 ..	100·0	100·0 ..	59·2 ,,

(The planimetric index indicates the percentage of the plane of a given nucleus as compared with the total sum of the nuclei of the same side. Example: The planimetric index of right caudate nucleus is $\frac{5{\cdot}760 \times 100}{21{\cdot}870}$.) This table indicates that the relative sizes of the different nuclei on the right or left sides are approximately the same, but all the nulcei on the left side are smaller than those on the right side. The left putamen is largely involved in this atrophic process, while the left pallidum I showed only a reduction of one-third of its plane as compared with the right side.

One section through the cerebellum was measured in order to determine the approximate degree of the deviation between the right and left sides. These are the perimeters which were found: Visible surface, right 12·5 cm., left 12 25 cm.; difference 3 per cent. Total surface, right 68·8 cm., left 61·7 cm.; difference 10 per cent. These differences are within normal limits and do not indicate an atrophy of one half of the cerebellum as compared with the other half.

Histological report.—Sections of the spinal cord stained by Loyez' method show a pallor of the inner half of the posterior columns at the level of the lower dorsal segments, with reduction in the number of myelin sheaths. The walls of the blood-vessels in this region are thickened and their lumina reduced.

Arteriosclerotic changes of medium degree are found in the pons, the ventral half of which was more involved.

There are no striking histological changes in the cerebellum. The nucleus ruber and substantia nigra on both sides are normal. The left body of Luys is richer in glial nuclei than the right, and its ganglion cells contain more pigment, which enlarges the cells and forms globule-like appendices. In Loyez-stained sections the inferior part of the capsule of the body does not stain as well as the rest: the fibres forming the tract which connects the outer part of pallidum II with the mid-brain nulcei (*faisceau pallidal de la pointe*) are less numerous than in the corresponding right sections. The lateral nucleus of the left thalamus is rich in glial cells, and its ganglion cells show different degrees of atrophy; they are more darkly stained by Nissl's method, and many of them are completely broken down. The dorsal and the medial nulcei of the left thalamus appear normal. The right globus pallidus shows no striking changes. In the left pallidum II there is scattered perivascular œdema with some round-cell infiltration, but the ganglion cells are not markedly affected.

A similar picture was seen in the left pallidum I. Towards the putamen there is an increase in the number of glial nuclei and in the number of diseased ganglion cells. In the striate body these histological changes reach their maximum, and the process of degeneration involves both large and small cells. In the oral part numerous gitter cells replace the broken down tissue, which is partly intermingled with connective-tissue fibres. Fat stains show numerous granules in the glial and ganglion cells. Iron stains reveal granular cells laden with iron pigment, and the left striate body and pallidum stain more bluish than the right side. Glial stains reveal a preponderance of microglia in the region of the left striatum in the proximity of the lesion. In sections stained by Loyez' method the right basal ganglia are coloured distinctly darker, and contain more myelinated fibres than the corresponding left nuclei (fig. 4).

Comment.

On comparing the five cases published previously with the present case, it appears that the hemichorea affected only persons older than 50 years of age (Jakob does not state the age of his patient), who were suffering from arteriosclerosis of the brain. In the cases of Vogt, Jakob, Austregesilo and Galotti, and Souques and Bertrand, and in the present case, the oral part of the caudate nucleus and putamen was destroyed by a cyst formation; in the case of Lloyd and Winkelmann the lesion was more occipital and did not affect the caudate nucleus. Unfortunately neither Vogt, Jakob or Austregesilo and Galotti give detailed histological reports which would allow a further comparison of the six cases. Lloyd and Winkelmann state that the lesion also involved the thalamus and the body of Luys on the right side. Besides, they found "a generally medullary fibre poverty in the striatum, particularly on the right, with

a generalized atrophy." In the case of Souques and Bertrand, the deeper layers of the corpus callosum were destroyed, as well as the anterior third of the posterior limb of the right internal capsule and the anterior peduncles of the right thalamus; pallidum I and the body of Luys were also affected.

In studying Plate XXVII, figs. 3 and 4, of Vogt's case [6], the left caudate nucleus and the visible part of the left putamen seem to be smaller than the corresponding right nuclei. In the present case there was a marked atrophy with poverty in myelinated fibres in the whole right caudate nucleus, putamen, both pallida and the body of Luys. It is evident that in Lloyd and Winkelmann's case, and in my own, this general atrophy of basal ganglia and the diminution of fibres cannot have been secondary to the lesion which produced the hemichorea, but must have preceded the extrapyramidal disturbance. In Lloyd and Winkelmann's case the choreiform movements appeared only three weeks before death, in the present case five months before. Further, in the cases of Lloyd and Winkelmann, Souques and Bertrand, the body of Luys was involved; in my case this nucleus was smaller than the right and richer in glial nuclei. The inferior part of its capsule was poorly developed, and the fibre tracts connecting the pallidum with the body of Luys and the substantia nigra on the left side showed a diminution of myelinated fibres. In these three cases part of the thalamus was also involved in the pathological process.

Localized lesions of the striate body are very frequently encountered without clinical signs of hemichorea. Jakob already emphasized the complexity of the problem, and pointed out that cases of localized lesions of the striate body which are combined with hemichorea are extremely rare compared with the relative frequency of such lesions found at autopsy. He thought that other pathological changes in the motor apparatus and "individual conditions" must be combined with such striate lesions in order to produce the choreic syndrome. He did not, however, express an opinion as to the nature of such accessory conditions.

The lesions known to produce hemichorea may occur, as stated at the beginning of this paper, in the anterior cerebellar peduncle, in the thalamus, in the neostriatum and in the body of Luys. It appears that lesions in any part of the sestructures may eliminate a path the integrity of which is necessary to the absence of chorea. Lesions of the cerebello-rubral system do not produce chorea, nor do lesions of the thalamo-cortical fibres. In the cases of Lloyd and Winkelmann, Souques and

Bertrand, and my own, the lesions involved at the same time the external nucleus of the thalamus, the anterior limb of the internal capsule, the striatum and the body of Luys.

Beyond describing the various parts of the path—anterior cerebellar peduncles, thalamus, striatum, body of Luys—and the lesions which were found in this case, no conclusions can be drawn, and it is felt that conclusions which state that the lesion which causes hemichorea must lie in a specific part of this path, are, at present at least, premature.

Conclusions.—Localized macroscopic vascular lesions of the striate body in adult people do not produce the clinical picture of hemichorea. They have to be associated with more generalized histological lesions of the striatum, the pallidum, thalamus, or the body of Luys. In the present case of right hemichorea, a cyst formation in the oral part of the left striate body was combined with hemiatrophy of the whole left striatum, the left globus pallidus, the left body of Luys. The histological picture showed increase in glial nuclei and poverty in myelinated fibres in the atrophic nuclei. There was also found sclerosis of ganglion cells in the lateral nucleus of the left thalamus.

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TRIGEMINAL NEURALGIC PAIN ASSOCIATED WITH MULTIPLE SCLEROSIS.[1]

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SEVERE protracted pain in cases of multiple sclerosis is so rare that it seems hardly worthy of discussion. However, during the course of a long series of cases of multiple sclerosis four were noted in which the patients complained bitterly of intense paroxysmal neuralgic pain localized in one or more divisions of the fifth nerve. To these unfortunates pain seemed to be the most important complaint in their disease, and they demanded relief regardless of what else could be done to ameliorate their general helplessness. Accordingly, these cases stand out in bold relief from the general run of cases of multiple sclerosis and as such demand some inquiry.

Similar cases have been described by competent observers and in at least one instance careful necropsy suggested causes for the pain. Oppenheim reported the case of a patient suffering from multiple sclerosis whose early yet permanent symptom was severe neuralgic pain in the area of the fifth nerve. The patient died, and at necropsy a plaque was found on the root of the fifth nerve at the zone of its entrance into the pons. A drawing of a microscopic section of the pons appears in Oppenheim's textbook of nervous diseases. Marburg found the same pathological changes in a similar clinical case, and Berger, in a study of 206 patients suffering from multiple sclerosis, noted in the case histories of two that severe paroxysmal trigeminal neuralgic pain had been present at some time during the course of the disease. In one of these cases the pain was the initial symptom and it had persisted from the beginning of the disease, a period of five years. Gullain noted the presence of neuralgic pain in the area supplied by the fifth nerve in two patients at the beginning of the disease. Unfortunately, the foregoing observers did not give enough detail with regard to the type of pain observed. In most instances it

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was described as neuralgic and paroxysmal, but the symptoms were not subjected to the minute scrutiny that modern clinical experience has demanded for the purpose of classification.

Report of Mayo Clinic Cases.

Case 1.—*Multiple sclerosis of twenty years' duration: Disease far advanced and all symptoms well marked: Paroxysmal trigeminal pain for six years in the left second divisional area without any objective sign of injury to the fifth nerve: Section of the posterior root: Complete relief from pain, but death four days after operation: Necropsy.*—A man, aged 44, was brought to the Mayo Clinic, April 15, 1923, complaining of paralysis of the limbs and pain in the left side of the face. Twenty years previously he had noticed that in running his lower limbs felt heavy and stiff, and that he raised them from the ground with difficulty. Later he noticed that his gait had become so unsteady that he staggered in walking. With periods of remission and exacerbation he gradually became worse, so that eleven years before examination he required assistance to walk. Finally he was unable to walk at all and for five years his lower limbs had been completely paralysed.

Thirteen years previous to examination the patient had noticed tingling in the right hand and it had become difficult for him to write. His hand was unsteady, awkward and clumsy, and for eight years he had been unable to feed himself. The left arm became affected about two years after the right, but never to the same extent, and he was still able to feed himself with it although it was unsteady and clumsy. Fifteen years previously incontinence of urine appeared; this progressed to the stage of complete loss of control. Bowel control had remained normal. Transitory diplopia, lasting two to three days at a time, had appeared eight years before.

Six years previously the patient had experienced, for the first time, sudden, sharp, stabbing, shooting pain in the left cheek, localized in a small area. These paroxysms lasted only thirty to sixty seconds. At first single paroxysms occurred, with freedom for from three weeks to several months. Later many paroxysms appeared close together and lasted for hours and the remissions became shorter. The pain was extremely severe; it usually came on in the morning and was initiated by talking, chewing or washing the face. The patient remained as quiet as possible during attacks lest a paroxysm occur. The pain remained unilateral, but, whereas at first it occurred in the left cheek only, later it occurred in the left side of the nose and left upper lip, but was confined to the area supplied by the second division of the fifth cranial nerve. A year before he came to the Clinic three deep alcohol injections had been given within two weeks. Relief lasting for two months followed. A series of three injections were then given, with relief for four months. Thereafter the pain had become more severe, the paroxysms more frequent and the area involved greater. Eleven days before examination an alcohol injection had been given without any result whatever. On the morning of examination an

attack lasting an hour occurred. Each paroxysm lasted half a minute and was repeated every few minutes.

Examination.—The patient was emaciated, bedridden and obviously suffering severely. Between attacks, however, he was euphoric, happy and cheerful out of proportion to his affliction. His intelligence was somewhat low, but he co-operated readily and efficiently in the examination as far as pain would permit. Horizontal and vertical nystagmus and weakness of conjugate movement of the eyes to the right was noted. Vision was not reduced and the optic discs seemed normal. There was spastic paralysis of both lower extremities and of the right upper extremity. The left upper extremity was weak and spastic also, but to a much less degree, although there was marked incoördination in the finger-to-nose test. All tendon reflexes were exaggerated and the plantar reflexes were in extension. Abdominal reflexes were absent. There was diffuse diminution of pain and thermal sensibility, variable in degree and extent, over both lower extremities and over the abdomen up to the eighth dorsal segment. Joint and vibration sensibilities were markedly reduced in the four extremities. Speech was scanning in character. There was no anæsthesia or motor paralysis in the area of the fifth nerve, but the slightest stimulus applied to the left cheek or left side of the nose was enough to induce a severe paroxysm of pain. Talking, eating or laughing also caused pain, and altogether the patient's general behaviour was similar to that of a patient suffering from major trigeminal neuralgia. The urine contained a great deal of albumin and pus and a considerable number of red blood-cells. The Wassermann tests on the blood and spinal fluid were negative. The Nonne test of the spinal fluid was positive; there was one small lymphocyte for each cubic millimetre, and the colloidal benzoin curve was 0000003333000000. There was an area of sloughing over the sacrum from decubitus.

Treatment.—Because of the patient's suffering, which interfered with the last remnant of enjoyment of life, section of the posterior root of the left fifth nerve was advised. April 20, 1923, under general anæsthesia, this operation was performed by Dr. Adson. The patient's immediate post-operative condition was good and he was free from pain for four days, when he suddenly became cyanotic with rapid, shallow breathing and feeble pulse. He died within an hour.

Necropsy.—Apart from the observations on the central nervous system, to be later discussed, the more important changes discovered were enormous dilatation of the stomach, chronic inflammation of the bladder with trabeculation and diverticula, moderate arteriosclerosis, emaciation and decubitus. The immediate cause of death was assumed to be acute dilatation of the stomach.

When the skull was opened gross signs of meningitis were not present. The operative field apparently was not infected and there was no more hæmorrhage than would be expected following such an operation. The posterior root of the left Gasserian ganglion had been completely severed close to the central end of the ganglion. The brain, spinal cord, and the right and left Gasserian ganglia with their posterior roots were removed, and also the peri-

pheral branches of the fifth nerve as far as it could be reached by the intracranial route. These tissues were immersed in formalin, and after they were satisfactorily fixed and hardened an endeavour was made to establish the diagnosis of the disease in the central nervous system, and to estimate, as far as possible, its anatomical distribution. The cerebral hemispheres were separated from the brain-stem by section through the upper part of the mesencephalon, and the cerebellum was detached by severing its peduncles. Coronal sections were made through the cerebral hemispheres. In the white matter in each section innumerable greyish plaques, varying from minute specks to 4 or 5 mm. in diameter, were clearly visible to the naked eye. These were scattered irregularly throughout the white matter, but were more numerous in the neighbourhood of the lateral ventricles. In the subependymal layer of the ventricles were translucent grey areas about 8 mm. thick, following closely the contour of the ventricular cavities and chiefly in the region of the posterior and inferior horns. Only a few plaques were found near the third ventricle and the cortical grey matter was comparatively free.

Microscopical examination of the isolated plaques and those confluent in the ventricular wall illustrated the following characteristics. With a modified Weigert myelin-sheath stain the plaques were completely demyelinized and stood out as white sharply outlined circular areas on a black background. Many of the plaques surrounded a central blood-vessel. With higher magnification the myelin sheaths appeared to end more or less abruptly in the margin of the plaques, with here and there a solitary nerve-fibre still retaining its stain crossing the dead white area. With Bielschowsky's and Orlando's stains the axis-cylinders in the plaques were relatively unaffected and continued on their course across the margin of the plaque into its centre. With Weigert's neuroglia stain there was an obvious increase in glial fibres, so there was a dense felting more marked toward the centre. Study of the central blood-vessel present in some of the smaller plaques showed some of the perivascular spaces filled with compound granular corpuscles, which when stained with Scharlach R. were found to be loaded with fat granules. Areas lining the walls of the lateral ventricles were merely a confluence of innumerable plaques with the characteristics described. Altogether the histology of the plaques was identical with that described in multiple sclerosis, and such was the final pathological diagnosis.

Sections were made of the spinal cord and medulla at various levels and stained with Weigert's myelin-sheath stain. Numerous plaques, scattered indiscriminately, were found along the course of the spinal cord. There was rather marked degeneration of the pyramidal tracts in the dorsal and lumbar regions. In the upper cervical spinal cord and lower medulla a plaque was found in the spinal root and nucleus of the fifth nerve on the right side (fig. 1). This, however, disappeared at a level corresponding to the upper third of the inferior olive.

Serial sections, 20 micra thick, were made of the pons throughout its extent and every fifth section was stained with Loyez' modification of Weigert

myelin-sheath stain. Sclerotic plaques of various sizes were found in every section, but the largest was that in the floor of the fourth ventricle, occupying nearly its whole length and breadth. Opposite the main sensory nucleus of the fifth nerve and on the right side there was a tongue-like projection of the plaque which enveloped this nucleus as well as the fibres of the

FIG. 1.—Section through the medulla in the region of the inferior olive showing plaque on the descending root and the nucleus of the right fifth. (×4·5.)

FIG. 2.—Section through the middle of the pons showing a large plaque in the region of the nuclei of the right fifth nerve and its descending roots. The floor of the fourth ventricle is occupied by a large plaque. Numerous larger and smaller plaques in other areas of the section. (×4·5.)

median fillet on that side. The nuclear cells, however, seemed intact although surrounded by sclerotic tissue. The left fifth nerve nucleus was intact (fig. 2).

Both roots of the fifth nerve were traced upward in their oblique, outward course within the pons; they emerged in the usual manner and formed the posterior root of the nerve. As traced upward each root took the myelin stain well and did not seem at all degenerated, but just at the point of emergence of the left root two plaques appeared and engulfed its fibres. On the right side the root seemed normal (fig. 3). Study of the left root in its extrapontine course showed that the sclerotic process extended outward a short distance within its substance, and then the root became seemingly normal again up to the point of its severance by the surgeon, at which point it was surrounded by a blood-clot. Study of the left Gasserian ganglion with various stains (Weigert myelin-sheath, van Gieson, hematoxylin and

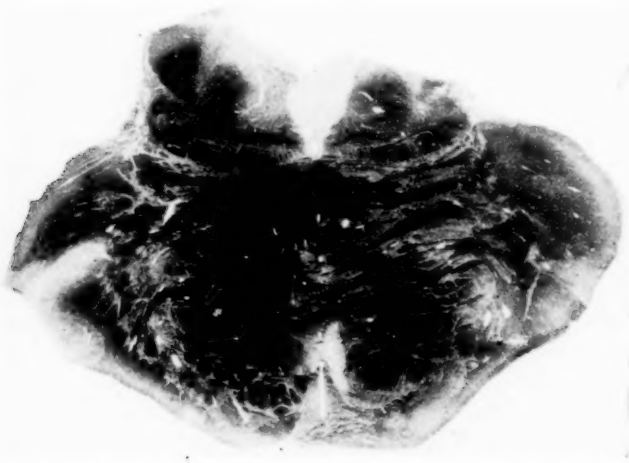

FIG. 3.—Section through the pons at the level of the emergence of the left trigeminal nerve root. Two plaques are to be seen invading the emerging fibres of the root. The right fifth nerve root at this level is normal. Numerous smaller plaques scattered throughout the section. (×4·5).

eosin, and Nissl's stain) showed nothing more than could be attributed directly to the trauma of surgical intervention. The ganglion was sectioned serially and scrutinized minutely for sclerotic plaques but none were found. Hæmorrhage was found on one surface, and a mild inflammatory reaction was present close to the hæmorrhagic area. The ganglion cells, however, were normal in size, the nuclei were central and the pericellular capsules intact. The blood-vessels were normal and unusual pigmentation was not present. The intraganglionic nerve-fibres took the myelin stain well, and neither the entering root-fibres nor the issuing nerve-trunks showed frank evidence of degeneration. The right Gasserian ganglion and corresponding posterior nerve-root were normal in all respects. Studies of the mid-brain

showed relatively few plaques, and the mesencephalic root of the fifth nerve on each side was apparently normal. The cerebellum was comparatively free, except for one large plaque about 2 mm. in diameter in its white matter near the nucleus dentatus and roof of the fourth ventricle.

Summary of necropsy findings.—The disease of the central nervous system from which the patient suffered was multiple sclerosis. Innumerable plaques were scattered throughout the brain and spinal cord. Well-marked and relatively old plaques were found on the main sensory nucleus of the right fifth nerve. On the right side and on the lowest part of the spinal nucleus and root was a plaque. The right posterior root was normal, but on the left side at the point of emergence of the root from the pons and for a short distance along its extrapontine course, there were two relatively old sclerotic plaques. The Gasserian ganglia and the central portions of the peripheral nerve-trunks were free from any sclerotic process.

Comment.—The case was one of typical slowly progressive, well-developed and well-established multiple sclerosis. Whatever doubts might exist with regard to the clinical diagnosis the observations at necropsy establish beyond question the nature of the disease. The condition had existed for twenty years and was readily recognized, but for six years pain had been present identical to that described under the synonyms of tic douloureux, trifacial neuralgia and major trigeminal neuralgia; none of the features of the latter disease was lacking. The pain was lancinating, paroxysmal and distinctly confined to the second divisional area of the fifth nerve. The paroxysms, occasionally spontaneous, usually were initiated by moving or touching the face or jaws. Accordingly, during certain attacks eating and drinking were avoided and the patient's nutrition suffered. The usual periods of remission had occurred, and on two occasions the injection of alcohol into the trunk or peripheral branches of the fifth nerve brought relief for a few months. In spite of the severity of the sclerotic process in the pons, no objective or subjective signs of injury to the fifth nerve were present, apart from the paroxysmal pain.

From a review of the clinical history of this one case, and without the necropsy data, it might be assumed that two distinct disease entities were present in one patient at one and the same time, namely, multiple sclerosis and trigeminal neuralgia. It could have been argued that the presence of a pain syndrome identical with trigeminal neuralgia in a case of multiple sclerosis was a mere coincidence of two unassociated diseases. However, the experience of this patient as well as that of three other patients similarly afflicted, tends to suggest some causal relationship between the multiple sclerosis on the one hand and the trigeminal pain on the other.

Case 2.—A nine-year history of multiple sclerosis with well-marked signs and symptoms; a three-year history of paroxysmal trigeminal pain in the second divisional area on the left side; no relief from alcohol injections into the infra-orbital nerve; section of posterior root of the fifth nerve and complete cessation of pain.—A woman, aged 39, first came to the Mayo Clinic, October 7, 1918. She had complained then of difficulty in walking. Thirteen years previous to this visit she had first experienced transitory diplopia. This recurred four years later and at this time also there appeared divergent strabismus of the left eye, formication and hyperæsthesia over the left side of the face, dropping of the left angle of the mouth, tinnitus in the left ear, and extreme vertigo with vomiting. These symptoms came on within two or three days and with fluctuations; they disappeared within a few months. Five years before examination weakness in the left lower extremity with a tendency to drag the limb in walking had developed. This reached its maximum in six months and then gradually disappeared. Two years previously this trouble had recurred, gradually developing within a space of two or three months. This time the patient did not recover but became worse, and the right lower extremity became affected also. Her gait had become stiff, slow and unsteady. She dragged her limbs as she walked, particularly the left side.

Examination.—The general physical examination showed a somewhat emaciated woman unable to walk without assistance. Ophthalmoscopic examination of the fundus showed slight pallor of the optic discs, more marked on the left side, but vision was $\frac{6}{15}$ in both eyes. Marked sustained nystagmus, both vertical and horizontal, was present. There was no facial weakness, but the left eye could not be closed without the right participating. There was general increase in tone, loss of speed, and weakness in all extremities. The weakness was slight in the upper limbs but reached the degree of a marked spastic paresis in the lower. All tendon reflexes were increased, more markedly on the left side. Babinski's sign was present on both sides. Abdominal reflexes were absent. There was marked movement tremor in the upper and lower extremities, and in walking there was a coarse, general tremor. If the patient attempted to stand with her eyes closed and feet close together she swayed and fell and the general tremor was again obvious. There was slight diminution of pain and thermal sensibilities over both extremities, but in all other areas, including that supplied by the fifth nerve, sensibility was normal. There was no involvement of the motor division of the fifth nerve. There was no disturbance of joint, vibratory or deep pain sensibility. The gait was spastic, more marked on the left side, with the general tremor mentioned. Speech was normal, and sphincteric control was normal. The Wassermann tests on the blood and spinal fluid were negative. The Nonne test on the spinal fluid was negative and there was only one small lymphocyte in each cubic millimetre. A diagnosis of multiple sclerosis was made. The patient was not seen again for six years.

The patient again visited the Clinic, August 21, 1924, because of paroxysmal pain in the left side of the face. Three years before this second visit

she had an attack of paroxysmal sharp shooting pain in the left cheek. All her teeth had been extracted and the pain disappeared for a year. It returned, however, and for two years preceding the second visit attacks had occurred periodically every few months until three months previously, when they remained without remission. The sudden, sharp, severe pain was confined to the left cheek and the upper and lower gums. It lasted only a few seconds and was accompanied by a burning sensation at the left angle of the mouth. The pain ceased between paroxysms. Many paroxysms were spontaneous, but most of them were initiated by touching or washing the left side of the face. Talking, chewing, laughing or drinking, or even opening the mouth, also brought on the paroxysms. The patient had been unable to eat a sufficient amount of food and therefore had lost weight. The pain was confined to the area supplied by the second and third divisions of the fifth nerve. The paroxysms came on every few minutes, and the frequency seemed to depend on physical activity, being further apart during immobility of the face, tongue and jaws. A week previous to the second visit alcohol had been injected into the left infra-orbital foramen and nerve, without relieving the pain. It produced, however, a numb sensation in the area supplied by this nerve; immediately prior to the injection there had been no paræsthesia in the painful area. So far as the multiple sclerosis was concerned the patient had not become much worse. Moreover, she had had a normal pregnancy and delivery during the period between the two visits. The child had apparently developed normally and was four years of age. In the meantime, however, the patient had been unable to walk without holding to objects and then only for a few yards.

Examination.—The patient was in poor general condition, weighing only 101 lb. as compared to her normal weight of 158 lb. She was obviously suffering extreme paroxysmal pain, and she refrained from even the slightest movement of face or jaws, fearing further suffering. Profuse lachrymation occurred during a paroxysm. Vision in the right eye was $\frac{6}{6}$ and in the left $\frac{6}{36}$. Ocular movement in all directions was slightly limited, and vertical and horizontal nystagmus was marked. There was pallor of the temporal part of the right optic disc, and all the signs of simple optic atrophy were present on the left side. There was slight weakness of the left lower lid and angle of the mouth, and well-defined hyperæsthesia to pain, tactile and thermal sensation over the left cheek, in the region of the infra-orbital foramen, presumably the result of the recent alcohol injection. The patient insisted that previous to the injection the sensation in the cheek had been normal. The symptoms noted at the neurological examination were the same as on the examination seven years previously, but further advanced. Spastic paraplegia was still present, and now the patient could not stand without support. The arms were spastic but not to the extent of the lower extremities. The movement tremor was more marked and the voice was slow and monotonous. The sphincters had remained intact.

Treatment.—A deep alcohol injection into the fifth nerve was given August 25, 1925; the second division was injected superficially August 27

and 28. Since these injections afforded but slight relief, regional and ether anæsthesia was administered on August 31, and the sensory portion of the posterior root of the left Gasserian ganglion was cut, leaving the motor-root intact. The patient's convalescence was uneventful and she was completely relieved from pain. The usual post-operative anæsthesia of the area of the fifth nerve was complete.

Two years after the operation the patient wrote that she was free from pain in the face and consequently had gained in weight. So far as the other difficulties were concerned she was still unsteady in her gait and required assistance to get around the house, but her general comfort and happiness were greatly increased.

Case 3.—A five-year history of multiple sclerosis with all signs well advanced, three and a half years of paroxysmal trigeminal pain, first in the third division later spreading to the second; section of the posterior root with complete relief for six years.—A man, aged 34, came to the Mayo Clinic April 1, 1919, complaining of difficulty in walking and pain in the right side of the face. He had been well up to three years previously, when he had noticed progressive unsteadiness of gait and weakness of the lower extremities. He staggered and stumbled when walking, and it had become more and more difficult to start urinary flow. Sexual power had diminished. For eighteen months he had suffered from attacks of pain in the lower jaw, lasting a few hours, then disappearing for several months. The pain was sudden, sharp and paroxysmal, and radiated along the ramus of the jaw into the right temporo-mandibular articulation. During attacks, talking, eating, drinking, cold draughts, or any touch on cheek or chin initiated a paroxysm. Therefore, he was scarcely able to eat, and subsisted on semi-solid foods that did not require chewing.

Examination.—The patient appeared to be fairly well nourished. He walked with a stiff and unsteady gait. The left optic disc was slightly pale. There was moderate spasticity of the lower extremities, and their tendon reflexes were exaggerated; ankle-clonus and Babinski's sign were present bilaterally. Abdominal reflexes were absent. Vibration and joint sensibility were disturbed in both lower extremities, but there was no change in pain, tactile or thermal sensibility. Incoordination was definite and marked in both lower extremities, and the gait was ataxic and spastic. At this time he could walk unaided. In the upper extremities also incoördination was definite, but more marked on the left. He swayed when he stood with both feet together and with his eyes closed, and he could not balance himself on one foot. The Wassermann tests on his spinal fluid and blood were negative. The Nonne test on his spinal fluid was positive, and there were fifteen small lymphocytes to the cubic millimetre.

The patient came to the Clinic again, August 17, 1921, two years after his first visit, and five years after the onset of the disease. He had become very much worse so far as his walking power was concerned. Paroxysms of pain were more severe and more frequent, and the pain had spread to involve the cheek and upper jaw as well as the lower jaw. Remissions between attacks

were shorter. He had been unable to keep up his nourishment and had lost considerable weight and strength.

The patient was undernourished and, whereas before there was only slight pallor of the left optic disc, it was now quite marked and there was loss of substance in the nerve-head, with signs of beginning simple optic atrophy. However, vision was not greatly reduced in either eye. He was then unable to walk without assistance and his legs were much more spastic and ataxic than they had been. Abdominal reflexes were still absent and ankle-clonus and Babinski's sign were even more marked. The left arm remained incoordinate but not more so than previously. There was now almost complete absence of vibration sensibility in both lower extremities, and the joint sensibility of the toes was more impaired. At no time were there any objective signs of injury to the fifth nerve. On the previous visit the patient could walk, now he was almost bedridden. Trouble with the bladder became more serious, and cystoscopic examination revealed a trabeculated bladder and cystitis. The Wassermann tests on the blood and spinal fluid were again negative, and as before the Nonne test was positive and the small lymphocytes had increased, this time being seventeen for each cubic millimetre. Two deep alcohol injections were given six days apart with only temporary relief for a few hours.

While the patient was under observation, the right arm in turn became weak and incoördinate, and nystagmus and a movement tremor of both upper extremities developed. In spite of this he was distinctly euphoric when he was not suffering, but the pain became the dominant feature of his illness. It was sudden, sharp and severe, and was released by the slightest stimulus applied to the areas supplied by the second and third divisions of the right fifth nerve. It was confined strictly to those areas and was always paroxysmal; it interfered with nutrition and sleep to such an extent that with his other troubles he seemed to be slipping away. It seemed advisable, therefore, to relieve his suffering by surgical means, even in the face of the diffuse central nervous system disease.

Treatment.—September 20, 1921, under general anæsthesia, Dr. Adson cut the posterior root of the right Gasserian ganglion. The patient was relieved from pain; he gained weight and his general condition improved. With the usual optimistic euphoric personality, freedom from pain seemed to be the patient's highest desideratum, even though everything else that made life worth living were lost forever. He left for home November 21. The last communication received from him was dated February 17, 1927, that is, six years after the operation. He had never had any of the old pain, but he was much more helpless and gradually losing the use of his arms also. All he could manage to do was to sit up in his wheel-chair; he could not turn over in bed or feed himself or even light his own pipe. On the other hand, he appeared well, ate well and enjoyed food, and complained little.

Case 4.—Multiple sclerosis proceeding with characteristic remissions and exacerbations for fourteen years; twelve years of paroxysmal pain in all three

divisions of the left fifth nerve. Later, similar but milder pain in the right side; relief with section of the left posterior root and deep alcohol injection of the right fifth nerve; seven years later the patient was free from pain on the left side, but pain on the right had returned.—A woman, aged 42, came to the Mayo Clinic, August 14, 1920, complaining of pain in both sides of the face and unsteady gait. Fourteen years previously, following childbirth, she had noticed numbness in the fingers and a sense of unsteadiness in gait. These symptoms fluctuated considerably at the onset, with periods of improvement followed by exacerbations. There was a general tendency to get worse, and during two years previous to her visit to the Clinic the unsteadiness of gait had become permanent and she was unable to walk without assistance or support. Twelve years before examination sudden sharp, lancinating pain had developed in the left lower jaw radiating to the left temporo-mandibular joint. This spread to include the left cheek, temple and forehead. The pain usually was paroxysmal, coming on in attacks and lasting from ten days to three weeks. The paroxysms lasted only for from thirty seconds to two minutes, and were initiated by peripheral stimuli, such as talking, laughing, chewing, or washing the face. The remissions became shorter so that at the time of examination the pain appeared almost daily in the entire area of the fifth nerve on the left side. Five to seven years previously the same type of pain had commenced in the right cheek and temple. It was infrequent, not as severe as that on the left side, and only occasionally was produced by washing the face.

Examination.—The patient was undernourished and suffering from severe intermittent and paroxysmal pain in the left cheek, temple, forehead and jaws, with complete freedom between paroxysms. She appeared to dread moving any part of the affected area. There was nystagmus in all directions and a markedly ataxic gait. The lower extremities were slightly spastic, and all tendon reflexes were exaggerated with ankle-clonus. Babinski's sign was present on both sides, and Oppenheim, Mendel-Bechterew, and Chaddock signs were also present; there was, however, no marked weakness of the limbs. Abdominal reflexes were absent. There was slight incoordination of the upper extremities and marked incoordination of the lower. Sensory changes were not manifest. The function of the fifth nerve was normal, apart from the pain and the reaction to superficial stimuli. Sphincteric control was normal. The Wassermann tests on blood and spinal fluid were negative, and in the spinal fluid the cell-count was one lymphocyte for each cubic millimetre.

Treatment.—On account of the patient's obvious suffering a deep alcohol injection of the fifth nerve on the right side was given on September 1. This relieved the pain on the right side completely. The posterior root of the Gasserian ganglion on the left side was severed by Dr. Adson under general anæsthesia on September 11. Complete relief from pain followed, and at the time of the patient's dismissal, September 25, she was free from all suffering. Her general strength and weight had consequently improved. Her gait and ataxia, of course, remained the same. She was last heard of, March 26, 1927, nearly seven years after the operation. The pain on the left side had not

returned, but on the right side, where alcohol had been injected, pain had returned two months after her dismissal. Apparently it was infrequent and not severe enough greatly to interfere with her comfort. The course of the multiple sclerosis had been up and down without at any time reaching the stage of causing complete helplessness. At the present time she is still able to get around, but, of course, with difficulty.

Discussion.

These four cases present many similarities, both in the course of the disease and in its salient features. In each case the disease was so well established that three out of the four patients had had symptoms for twenty, nineteen and thirteen years respectively. The disease had begun, as it usually does, in early adult life. In the remaining case the onset had occurred at least five years prior to operation. In all, therefore, the signs of the disease were well-marked and established the diagnosis of multiple sclerosis beyond any reasonable doubt. It is of interest that two of the patients were observed over a period of years during the course of the disease. In all four the course had been the usual one of remissions and exacerbations. A complete physical examination was made in each case, including a routine neurological examination, spinal puncture and laboratory tests of the blood and spinal fluid. Possibly the unusual features of the disease led to a more careful and cautious appraisal of each case. Finally, necropsy was possible in one case in which the patient died following operation, and the conditions noted in the central nervous system after death were in accord with the previous clinical diagnosis of multiple sclerosis.

The pain syndrome in all four cases was identical, and without exception it followed the course of that in major trigeminal neuralgia, with the usual remissions and exacerbations of the latter. In three of the cases the pain was unilateral, in one both sides were affected. In Case 4 the three divisional areas were affected, in Cases 1 and 3 the second and third divisions were affected, and in Case 2 the second division alone. In the cases in which more than one division was affected the pain had commenced in the second or third division and spread to the other in the course of time. The patients' sleep was not greatly disturbed, but they dreaded the periods of activity enjoined by their daily needs and preferred to remain immobile lest an agonizing paroxysm be initiated. Periods of freedom from pain were common but they grew progressively shorter, and patients were ready to accept the operation of section of the posterior root of the fifth nerve as a

source of palliative relief, even though it brought no hope of cure of other complaints.

The relation of the period of pain to the whole course of the disease varied in the four patients; two patients, who had symptoms of multiple selerosis for nearly twenty years, suffered from pain only in the last few years. Two patients had had pain almost from the onset of the disease. One of the latter had had symptoms of multiple sclerosis for fourteen years and during twelve of these had suffered recurring attacks of paroxysmal trigeminal pain.

The resemblance of the pain syndrome in these cases to that in trifacial neuralgia led to the same surgical measures being advised as in the latter disease. So far as relief from pain is concerned, the operations were uniformly successful, since the three surviving patients have been free from pain to the present time, representing periods of two, six and seven years respectively. Alcohol injection of the nerve afforded temporary relief in a few instances, but to a much less degree. The justification for the major operation on these unfortunate patients is shown in their letters, wherein they all expressed gratitude for their relief, even though their other symptoms remain and their existence is necessarily crippled and disabled.

Pain in various parts of the body during the course of multiple sclerosis has been described by numerous authors, notably Oppenheim, Nonne, Berger, Müller, Barré, Schlesinger, Taylor, Keschner and Malamud, and Sittig. It is their common belief that pain is a somewhat rare phenomenon, but nevertheless occurs in a small percentage of cases. It seldom dominates the clinical picture, yet it may be the initial symptom and persist almost throughout the course of the disease. The pain is described variously as burning, lancinating, fulgurating, shooting, neuralgic, squeezing and lightning. Occasionally it may be severe and persistent. Müller has used the term *sclerosis multiplex dolorosa* for some of these cases Certain observers believe that the pain originates in plaques in the posterior columns, but a greater number believe that it originates in the posterior roots. However, the correlation of symptoms in multiple sclerosis during life with the pathological changes after death has been generally recognized as a difficult task. Taylor in particular has emphasized this; in one of his cases in which severe pain has occurred in the lower extremities it might have been expected that involvement of the dorsal nerve-roots would have been observed at necropsy, but unhappily such involvement could not be demonstrated microscopically. The reverse holds also, in that marked involvement of

areas of the neural axis by the sclerotic plaques may have no clinical counterpart during life. Oppenheim had found plaques continually in relation to the fifth nerve nucleus without disturbance in the sensibility of the face; on the other hand, he has assumed that the sclerotic plaque on the fifth nerve-root was the cause of the severe neuralgic pain from which his patient had suffered. It is therefore only with caution that any assumption can be made as to the possibility of one or the other sclerotic lesion being the sole cause of the symptoms in a given case. My experience in the one case which came to necropsy was identical with that of Oppenheim as to its clinical and pathological features. It is a temptation to regard, as he did, this lesion on the posterior root as the source of the patient's pain, especially when it is remembered that so far as could be ascertained the Gasserian ganglion and peripheral branches of the fifth nerve were normal. In this regard the question arises as to whether this entrance zone of the posterior root into the pons is a particularly dolorogenetic area. Certainly the posterior roots in the spinal canal are extremely sensitive, as any surgeon can vouch for who has extracted a particularly large cord tumour while the patient was under a local anæsthetic. Touching or manipulating the roots when they are insufficiently infiltrated with the anæsthetic produces sudden darting, lightning-like excruciating pain.

Again, there is ample pathological evidence to show that compression or irritation of the root of the fifth nerve by tumours of the cerebello-pontine angle will produce intense paroxysmal neuralgic pain in the face and jaws. The cases of Lexen, Krause, and Weisenberg are good examples of this, as is a similar experience observed by me, wherein the pain produced by a tumour of the cerebello-pontine angle was identical with that associated with tic douloureux. It is also worth while to mention the former theory of Redlich and Obersteiner as to the initial lesion in tabes dorsalis of the central nervous system. This was assumed to be at the point of entrance of posterior root-fibres into the dorsal columns of the spinal cord and at the point where they lose their neurilemmal sheaths. One of the earliest symptoms of tabes is the characteristic, sudden, paroxysmal, sharply localized lightning-like pain that may appear in the face as well as in the more usual site, the lower extremities.

Any discussion as to the site and mechanism of the pain in the four cases recorded here can be made only in a spirit of pure speculation. It is manifestly impossible to form conclusions that are convincing and free from criticism since there are so many possibilities of error. I

have attempted, therefore, only to demonstrate objective data, and my interpretation of these data should be accepted in the spirit in which they are offered. My observations of these four patients may be of some interest to those concerned in the problem of the causation of pain in general, and of that referred to the fifth nerve in particular. Our clinical experience is so far ahead of our knowledge of structural change that the mechanism of the production of many well-recognized clinical syndromes lacks explanation. Pain in particular is a purely subjective phenomenon, and presents some of our most baffling problems. My experience as well as that of others seem to establish the fact that it may occur in the trigeminal area as one of the rare symptoms of multiple sclerosis, but the essential mechanism must be studied further.

Conclusions.

(1) Patients suffering from multiple sclerosis may experience trigeminal neuralgic pain. While this symptom is comparatively rare in the disease, when present it may dominate the subjective clinical picture, and may appear early.

(2) The pain may resemble closely the type complained of in cases of major trigeminal neuralgia, and it is relieved by the same surgical procedures.

(3) Four cases of well-advanced multiple sclerosis and persistent paroxysmal trigeminal neuralgic pain were studied. One patient died, and at the necropsy the diagnosis of multiple sclerosis and the distribution of the disease were established.

(4) Plaques were found on the entrance zone of the posterior root in the pons on the side affected by pain. Smaller plaques were found on the spinal trigeminal root of the opposite side in the lower part of the medulla and upper part of the cervical spinal cord. The main sensory nucleus on the opposite side was also affected, but no apparent symptoms were produced. Both Gasserian ganglia were free from sclerotic processes.

(5) The interest in these cases centres equally on the mechanism of production of paroxysmal trigeminal pain, and the fact that this type may occur in cases of multiple sclerosis.

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ADDENDUM.

After the proofs of this paper had been corrected and it was ready for press, the article of Dr. Wilfred Harris, in vol. 50, p. 403, of this Journal appeared. He mentions having seen twenty-three cases of trigeminal neuralgia associated with chronic spastic paraplegia. He considered that the majority of these patients were in all probability suffering from multiple sclerosis. In several of these patients neuralgia was bilateral. The neuralgic pain was of exactly the same type as that seen in ordinary tic douloureux or paroxysmal trigeminal neuralgia. There was no objective evidence of damage to the fifth nerve.

THE NARCOLEPSIES.[1]

BY S. A. KINNIER WILSON.

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Introduction.

In recent years attention has been directed in increasing degree to the problems presented by disorders of the function of sleep. For this, neurology has been influenced largely though in no wise solely by the world-spread of epidemic encephalitis since 1918—a morbid condition characterized in a great majority of instances by disturbance of that function. Yet interest in questions bearing on the physiology and pathology of sleep is far indeed from being a new development in neurological science. So much is this the case that not a few recent communications dealing with the subject are reduced in value by ignorance or neglect of previous work of the same kind, or along the same lines; clinical syndromes are described as new when as a fact they

[1] A paper read at the Annual Congress of the Association of Physicians, Belfast, June 3, 1927.

are very old, and hypotheses are being evolved which suffer materially, even fatally, from failure to take all the germane data into consideration.

No one with any knowledge of the facts will be inclined to deny that this is patently the case in respect of what has been called narcolepsy —a term that has always been loosely employed, various writers having described clinical examples of sleep disorder under this heading without staying to define the sense in which it is being used. As a consequence, current ideas are uncommonly fluctuating where narcolepsy is concerned, though, as we shall see, attempts at delimitation have recently been made. A legitimate matter for discussion is whether, in point of fact, clinical instances of disorder of sleep are now occurring with greater frequency than in other times. This is evidently the opinion of Brailovsky [5], who says: "In der Literatur der letzten 3 Jahre finden wir ebenso viele Arbeiten über dieses Thema wie in den vorhergehenden 20 Jahren zusammen. Sicher steht das in Zusammenhang mit der von uns durchlebten Zeit." The statement, however, is inaccurate; long before epidemic encephalitis appeared many investigators had interested themselves in narcolepsy and cognate problems of sleep, as any one who makes himself familiar with the older literature (which is far indeed from being scanty) can readily realize.

My own experience in this respect may be given a passing allusion. I was house physician to the late Sir William Gowers at the time (1907) he wrote his fine little volume on "The Borderland of Epilepsy," included in which is a chapter devoted to some sleep symptoms—among them, so-called narcolepsy. I became familiar at that time with the subject and was on the constant look-out for clinical illustrations; yet in twenty years I saw no clear and impressive example (with one or two minor exceptions), until in the course of the last few months I have at length met with four striking instances of the former, the details of which are duly recounted below. Spiller [82] appears to have had an analogous experience; writing in 1926 he says: "A few years ago it was exceptional for a neurologist even with a large experience to see a case of typical narcolepsy. I have been impressed by the fact that at least three cases have come under my observation within a year." Were one to argue from these considerations a case might be made out for an actual increase in disordered conditions of sleep somehow related to the times in which we are living, and their ascription to processes arising out of epidemic encephalitis might be plausibly offered as one explanation, but at present I am far indeed from being convinced of any frequent ætiological association of the latter with

the former. Whether any apparent increase can be accounted for, to some extent, by augmented attention on the part of clinicians is a speculation not easily to be substantiated one way or the other; and as for the actual question of numbers of cases it also cannot be decided in any positive sense.

At the outset it appears desirable to discuss certain principles of a general kind which are highly important for the understanding of sleep problems.

Nosology.

From the viewpoint of nosology it is unscientific and reactionary to allude to any of the disorders of sleep as *diseases*. In particular, to describe the variety called narcolepsy as a *morbus sui generis* is a nosological error, yet in the communications of Henneberg [36], Redlich [70], Adie [1] and others this regrettably occurs *passim*. That a special *clinical* type of sleep disorder, a clinical syndrome corresponding to Gélineau's [28] original description of a "neurosis" to which he gave the name "narcolepsy" exists, may or may not be regarded as established; but, at the most, it remains only a clinical combination. The problem here is precisely analogous to that of epilepsy, which cannot be accounted a disease *per se*; fifty years ago Hughlings Jackson was pleading for recognition of "the epilepsies" as physiological, not pathological, conditions; epilepsy is a mere symptom, and idiopathic epilepsy is epilepsy of as yet unknown origin. Similarly, the morbid or diseased conditions of which narcolepsy is nothing else than the clinical manifestation still escape us in some cases, no doubt, but there can be no narcolepsy of any kind without a cause. No one has put the matter more clearly than Lhermitte [45], as the following citation shows: "Admettre la nature névrosique de la narcolepsie, c'est admettre par le fait même qu'elle est une maladie au vrai sens du mot, c'est-à-dire un état pathologique lié à la même cause et ressortissant à la même thérapeutique. L'énoncé même des caractères essentiels d'une 'maladie' suffit à montrer que la narcolepsie ne peut à aucun titre être assimilée à une affection autonome." And even if we take the particular syndrome of narcolepsy in the sense understood by Gélineau we shall see that recently more than one case has been reported in which it has appeared as a sequel to an attack of epidemic encephalitis, rendering its isolation as a *morbus sui generis* more impracticable even than before.

Terminology.

As already remarked, the term "narcolepsy" has been employed for more than forty years (Gélineau's paper having been published in 1880) in the vaguest way to signify any clinical variety of paroxysmal diurnal sleep, in recurring attacks of variable duration, or, indeed, of prolonged sleep states, the association of which with such known conditions as hysteria or other psychoneurosis or psychosis is unmistakable. Let it be noted that Gélineau's [28] definition—"Une névrose rare, caractérisée par un besoin subit, irrésistible, de dormir, ordinairement de courte durée, se produisant à des intervalles plus ou moins rapprochés et obligeant le sujet à tomber ou à s'étendre pour lui obéir"—is not of itself calculated to delimit the conception, as some recent authors have assumed, hence the vicissitudes which that conception has undergone, and which justified Lhermitte amply in alluding to "the narcolepsies," an expression which I myself consider eminently reasonable. Gowers [32] was one of the first to desiderate restriction of the expression, though recent writers have neglected his work; he pointed out that cases of *continuous* sleep should have another designation, as also those cases in which a sleep state is interrupted only when the sufferer is roused, for which he rather favoured the discarded term "somnosis." His conclusion was that narcolepsy should be used solely for cases in which "definite brief sleep interrupts a normal state." Naturally enough, it is impossible to say where the precise line is to be drawn between short and prolonged sleep (useful though the distinction is), and, for that matter, between sleep and lethargy, or trance; a recurrent tendency often characterizes trance-like states, while the question of whether the patient wakes spontaneously or only if roused cannot be utilized as a diagnostic criterion, since both conditions are frequently seen in one and the same case at different times. The truth is that all these distinctions are highly artificial, a consideration stressed recently also by Worster-Drought [96], who follows others in using the term solely in a symptomatic sense, to include all states of recurring diurnal sleep, whatever the ætiology.

A tendency, however, has revealed itself since the publication of Loewenfeld's [47] paper on the subject in 1902 to regard the narcolepsy of Gélineau as a special form constituted by a double symptomatology, viz., the combination of attacks of sleep with a peculiar proneness to the development of a transient condition of physical powerlessness under the influence of emotion, so that the patient's knees give way and he

may sink to the ground, without any loss of consciousness ("cataplexy"). This interesting phenomenon was noted by Gélineau's patient to occur when he laughed heartily, was observed also in Loewenfeld's case, and was raised by the latter author to the level of a diagnostic feature of narcolepsy *sensu strictiori*. Following him, Henneberg [36], Redlich [68], Stöcker [88], Kahler [42], Jolly [41], Singer [77], Münzer [59], and some other German authors consider the combination pathognomonic of narcolepsy, and would reserve the use of the term for such cases only, a line which is also strongly advocated by Adie [1] in his recent able thesis on the subject. On the other hand, Goldflam [30], Curschmann and Prange [15], and others hesitate to regard attacks of powerlessness as essential to the conception; Matzdorf [52] is convinced of the occurrence of rudimentary and atypical cases which do not correspond *in toto* to the above-mentioned combined form, and in this respect is followed by Stiefler [87] in his latest paper. Along with a typical case two others characterized only by sleep attacks are included by Janzen [40] under the same designation, while a similar attitude is adopted by Adie, who accepts a case of his own, and two reported by Goldflam, in which no powerless phenomena were observed, as belonging to "true narcolepsy,' and says in so many words that "sleep attacks alone may occur in genuine cases."

Experience should surely have convinced the neurologist that no good purpose is served by a hard-and-fast schematization of a clinical syndrome—which is all that narcolepsy can possibly be—to fit a conception based solely on an original description; as we have just seen, some previous writers are compelled to widen the horizon of Gélineau's narcolepsy, a procedure which in my opinion is desirable and inevitable. The four personal cases detailed below belong clearly to the combined category of sleep plus emotional ("cataplectic") attacks, but my contention is that this category is nothing else than a special variety of narcolepsy, and that the latter expression, if it is to be employed at all, can have nothing else than a purely symptomatic and non-committal significance, and under no circumstances can be used for a "disease."

This consideration leads to a further point in terminology. The Greek word *νάρκη* signifies, according to Liddell and Scott, "numbness, deadness (Lat., torpor), caused by palsy, frost, fright, &c.," whereas the characteristic of the clinical states that here concern us is their *apparent* resemblance to natural sleep. Hence some think a preferable term is "hypnolepsy"—an "attack of sleep"—and it is interesting to note that this suggestion had already been made, not

originally by Singer [77] (as he, in ignorance of previous work, appears to think), but by Foot [25], and that as long ago as 1886. It was also advocated by Carl Camp [9] twenty years ago (1907). But "narcolepsy" has been given so long a start that it will perhaps now scarcely be overtaken.

The most recent terminological suggestions come from Wenderowic 92], who proposes "hypnolepsia genuina" for the Gélineau combined type, "hypnolepsia symptomatica" for the same type appearing as a sequel to, or in association with, a known pathological condition, and "narcolepsia" for symptomatic hypnoid conditions arising in the course of various diseases. I do not feel inclined to adopt this rather elaborate and unnecessarily dichotomizing classification, and conclude by repeating that for the purposes of this communication "the narcolepsies" stand for hypersomnic varieties of sleep disorder, whatever their ætiology, among which it is convenient to distinguish a combined type corresponding to that so well differentiated by Gélineau.

Personal Cases.

Case 1.—*Male case. Duration, three years. Cause unknown. Typical diurnal seizures of "natural" sleep of brief period, associated with typical attacks of tonelessness in limbs under the influence of emotion. Additional symptom, frequent attacks of diplopia.*

P. O. R., aged 24. Occupation, railway carpenter: unmarried.

History of present illness.—In April, 1924, the patient had his first attack of irresistible sleep when actually on the top of a signal post executing a repair. He managed to climb down and fell fast asleep at the foot of it, for perhaps half an hour. Ever since, he has suffered from continually recurring diurnal attacks of sleep, not a single day passing without them. About the same time as the original attack, or rather later, he noticed occasional double vision, independent of the sleep "turns," the images being on the same horizontal level and a little way apart. At a period subsequent to April, 1924, but not determined with exactitude, he noticed that whenever he laughed heartily he suddenly experienced a giving way of the knees, so much so that on several occasions he sank to the ground powerless, but was up again a few seconds later. Apart from this triad of symptoms he has no other complaint whatever. He has continued at his work.

Previous history.—Two years previously, playing football, he met with an accident and was badly concussed, but the skull was not fractured. Venereal disease has never been contracted.

Family history.—Nil ad rem.

Condition on examination.—The nervous system is normal in all respects, no trace of any organic condition being discoverable. There is no ocular paresis. Optic discs clear. Reflexes normal. The patient has not put on weight, weighing at present 12 st 4 lb., the same as at the outset. His facies reveals a state of mind of some anxiety in respect of emotional influences; rather frowning and somewhat subdued, he preserves a forced gravity. "I don't laugh any more than I can help" (see fig. 1).

Wassermann test in the serum, negative. X-ray picture of the sella turcica and vicinity, normal. Blood-sugar estimation (Dr. O'Flynn), normal curve.

FIG. 1.—Facies. Case 1.

This case conforms entirely to the syndrome of the first division of the first group (see below), presenting the combination of attacks of sleep and of "cataplexy." The former are always of brief duration, rarely exceeding ten minutes. From them the patient either awakes spontaneously, or can be at once aroused by a touch; he immediately feels fresh and continues his job. There is no background of continuous sleepiness, and no conscious feeling of physical tiredness or fatigue. The patient has slept many times while waiting his turn to come into my out-patient clinic, yet he never sleeps once he has entered, however long the examination. Personal observation of the sleep shows it to be seemingly a state of natural slumber, with no abnormal manifestation. At night, he passes some eight hours normally in sleep and has no

difficulty in awaking. In regard to the "powerless" phenomena, the sole excitant appears to be laughing, no other modality of emotional stimulus precipitating them.

Case 2.—Male case. Duration, nineteen months. Cause unknown. A brother suffers from some variety of so-called epilepsy. Typical diurnal attacks of sleep, and numerous attacks of cataplexy, especially with laughing but also under the influence of other emotions. If the patient forcibly keeps himself awake an attack of powerlessness will supervene instead of the sleep. Neurological examination during one of these reveals temporary complete loss of deep reflexes and an extensor response. Additional symptom, increase of weight, amounting to 2 st. since onset.

E. C., aged 26. Occupation, bricklayer: married.

History of present illness.—One Saturday evening, nineteen months ago, the patient was handed a paper informing him that he had won £5 in a football competition sweepstake. At this unexpected news, in the very moment of excited rejoicing, he suddenly felt helpless, his head dropped down, his limbs gave way under him and he "flopped" to the ground in a state of complete powerlessness. His head was sunk on his chest and he remained motionless, unable to move a finger, for about a minute. During the whole of the time he was entirely conscious of what was going on round about him. Immediately on recovery he felt perfectly well.

A short time afterwards (exact interval not known) he had his first attack of sleep, the circumstances of which are not remembered.

Ever since, he has had attacks both of sleep and of loss of power in the limbs under the influence of any emotion, the former occurring every day, often many times a day.

(1) *Sleep attacks.*—These are apt to occur particularly during his work, that of bricklaying. He often finds it monotonous, laying one brick after another, and the attack is usually ushered in by his mind wandering off his job: "I feel so used to it that my mind wanders off, where, I cannot tell, and then I drop off." The duration of the sleep is from three to four minutes: it is not preceded by any feeling of physical tiredness, but rather by this sort of "day-dreaming." Whenever he awakes he feels quite fresh. He either awakes spontaneously or is easily roused by a touch or a shake, but during the attacks he is soundly asleep, completely oblivious of his surroundings, and as a rule not dreaming. He has often fallen asleep at meals, "with my fork half way to my mouth." On the whole, he is as liable to fall asleep in the evenings at home as at work, yet he is confident that the warmth of the room, the time and place, make not the slightest difference, nor does the matter of interestedness arise. "I fall asleep at the pictures, or during a business conversation, just the same."

(2) *Attacks of loss of power and tone.*—The attacks of "tonelessness" arise in two distinct ways, either under the influence of emotion, or *because he is trying to avoid an oncoming sleep attack.* In regard to the first of these, any excitement suffices. Thus: "At a football match my legs give way just when some one is about to score a goal." Temper will bring on an attack, and it is particularly noteworthy that the tendency to it is always aggravated if the patient thinks that people are looking at him. "If I am playing the fool with the baby and my wife looks on and 'chips in,' I am sure to have an attack."

FIG. 2.—Facies. Case 2.

Laughing heartily never fails to bring on one of these "turns." It is equally remarkable that there is sometimes a relation between the emotion and the part of the body involved for the moment in the particular action concerned. Thus: "If I go to throw a stone at a glass bottle and any of the fellows are looking, the power seems to go out of my arm." "If I go to shoot the ball at goal and I see the fellows watching me the use goes right out of my leg; I simply can't do it."

According to the patient's own description, in these seizures the head falls forward on the chest in a nodding movement, the limbs are absolutely useless and powerless, the whole thing lasts a minute or two, and immediately after he is "as right as rain" and has no desire to sleep for the next hour.

As for the second causation, he has for long observed that *if he endeavours to keep himself awake forcibly, when sleep is imminent, an attack of powerless-*

ness will supervene, unaccompanied by any conscious emotion (unless we are to assume that the effort to avoid sleep is itself coloured with emotional tone). In the event, one or other attack is certain, though there have been occasions when deliberate shrugging of the shoulders has seemed to prevent it.

Of the greatest interest is the fact that when he has been asleep and dreaming, *the emotional content of the dream has precipitated an attack of powerlessness*. The worst he has ever experienced lasted for about a quarter of an hour, and occurred under the following circumstances. He had fallen asleep and was dreaming of a murder. The emotion experienced in association therewith brought on one of the "loss of power" attacks, so that he at once awoke and was fully conscious but was utterly unable to move a single finger. He thought he heard his brother and sister coming up the stairs; he tried to call to them but could not make a single sound; the more he tried the more intense became his emotion and the more absolute his helplessness; he lay thus, flat on the floor, motionless but suffering acute mental distress, for some fifteen minutes ere the attack dissolved itself spontaneously. Similar but less severe attacks have occurred when he has been dreaming of "terrible happenings" on other occasions.

Ordinarily, the patient's night sleep is of normal duration, and he wakes readily enough in the mornings.

At the commencement of his illness—which has never interfered with his daily work—he weighed 10 st. 6 lb.; he now weighs 13 st. (see fig. 2).

Previous history.—He has had no previous illness of any import, and no head injury.

Family history.—An elder brother, aged 32, suffers from some kind of fits. These began in 1919 and he has had only three in all. From the description given it is difficult to determine their exact nature; it appears he falls unconscious and is stiff in his limbs, but there is no change of colour, tongue biting, or involuntary micturition. As he served in the army during the war and saw much fighting, it is possible they have a psychogenic basis. Otherwise, the family history is negative.

Condition on examination.—The nervous system is normal in all respects. The reflexes are entirely normal, both cutaneous and deep. There is no ocular defect; the discs are clear. Wassermann reaction in the serum, negative. X-ray examination of the sella turcica shows it to be if anything on the small side. Blood sugar curve (Dr. Lawrence), normal in character.

The clinical value of this striking case of combined narcoleptic and emotional attacks is enhanced by the fact that I was able to observe one of the latter from beginning to end and to examine the patient's neurological condition during it. Dr. MacDonald Critchley, the present Registrar at the National Hospital, was with me at the time and helped materially in the examination, which was of necessity hurried.

Description of attack of "powerlessness."—While I was occupied

with another patient, E. C. was sitting behind me and, as afterwards appeared, was endeavouring to keep himself from falling asleep. Suddenly I heard a slight groaning sound coming from him, and at once looking round I saw his head nodding gently on his chest as he sank forward into a bent position and a moment or two later slid or "slithered" off the chair to the floor. The arms were by the side and the fingers semiflexed; the eyes were closed. Lifting the arms I found them absolutely atonic and flaccid; when let go they fell like lumps of lead by the side. The legs similarly were absolutely atonic, sprawling out on the floor. Lifting the eyelids I observed that the pupils reacted to light slightly but definitely, while the eyelid muscles were so toneless that little or no reflex contraction took place on my touching the cornea. Testing the knee-jerks I found them *completely abolished* on both sides. In the meantime Dr. Critchley had hurriedly taken off the patient's shoe and sock on the left side, and testing the plantar reflex obtained a slight but left definite *extensor response*, which he demonstrated to me and which I corroborated.

Just as we were finishing this rapid examination the patient suddenly said, "I'm all right, sir," in his ordinary voice, and with a faint smile. Muscular power came back; he moved his limbs, got on to the chair, and told us *he had been conscious the whole time*. He described what we had done, from lifting the eyelids to scratching the sole of the left foot. The duration of the whole attack was from sixty to ninety seconds. Testing his knee-jerks again, I found them *active*, even brisk, on both sides, two minutes after he had recovered.

So far as I am aware this is the first time a neurological examination of a patient in the cataplectic state has been recorded. When Adie's paper was published (September, 1926) it was therein specifically remarked that "no patient has been examined by a competent neurologist while in the cataplectic state."

I shall refer at a later stage to the significance of this observation, supplying as it does details of much importance for the study of the mechanisms involved and for the problem of the nature of the emotional attacks and their relation to the narcoleptic phenomena.

Case 3.—Male case. Duration, twenty years. Cause, unknown. There are three separable types of attack in this case; (1) *typical short diurnal sleep seizures;* (2) *typical (momentary) attacks of loss of power in the limbs under the influence of any excitement, laughter or*

rage; trance-like states of "absolute paralysis" during which no movement is possible, while consciousness is quite intact, brought on not by emotion but usually when patient is resting or sitting quietly, sometimes when on his feet.

T. T., aged 41. Occupation, boilerman: married.

History of present illness.—(1) As far as the patient is able to recall, he first noticed the tendency to *attacks of sleep* when he was in the Army in India, some twenty years ago. Scarcely a day has passed since then without his having fallen asleep, not once but a number of times. Without any preliminary warning in the shape of tiredness or sleepiness he suddenly falls "dead asleep"—a dreamless slumber of some four or five minutes' duration, from which he wakes spontaneously, or is aroused with ease and promptness. He has had "dozens" in the day. He has fallen asleep on his feet, and on one occasion injured himself by knocking against a lamp-post. He has had such attacks at his work, during meals, under every variety of circumstance, and cannot assign them to any special cause, such as monotony of occupation, warmth, time of year, diversion of interest, &c. As soon as the sleep is over he feels fresh and well, and makes no complaint of physical fatigue or of any general sleepiness.

(2) In addition, and for about the same period, the patient has noticed that any *emotional excitement causes a peculiar sensation of weakness in the limbs*, which results in their giving way, so that on numerous occasions he has sunk down on his knees, or actually fallen. For instance, on one occasion he saw a horse bolt and without further warning his knees weakened under him and he went down. Its duration is, however, exceedingly brief, "almost momentary," and yet there is time for him to be well aware of his heart "beating like anything," and of a very definite feeling of tingling over the head, cheeks, and body, "like a lot of strings moving over my face." "For the moment I feel as if my blood were clotting." Laughter brings on these attacks of loss of muscular power, as well as annoyance or anger. In his own words: "If I were to try to strike one of my children it would come straight over me before I could do so, and I would go flat on the floor." This has actually happened on more than one occasion.

(3) The third type of attack is as follows: at any time, usually when sitting quietly in a chair, lying on a sofa, leaning against a wall, and so on, there will suddenly steal over him a "paralysing feeling": his eyes close but he never loses consciousness and is acutely aware of what is going on, his limbs relax and while the attack lasts he is *absolutely incapable of moving* a finger tip through a fraction of an inch. "I am paralysed from head to foot; some one could murder me and I could not move." On more than one occasion he has had a cigarette in his hand when this type of attack came on, and has actually burnt his fingers badly without being able to make a single movement to let it drop. The patient says that he "fights against it" inwardly by every means he can think of, yet not a sign of this is visible to the looker-on, who sees him

lying or sitting motionless. It appears that he eventually is able to make a grunting noise in his throat to attract the attention of his wife or other person; if he is touched or shaken forcibly the spell is at once broken and he recovers instantaneously. During the attack he is unaware of any feeling of fear, though he can neither move nor speak, but he is frequently conscious of his heart beating, "as if it were bursting." The duration of the attack is usually from three to five minutes.

He made spontaneously the interesting statement that "if I was to try and prevent myself from a sleeping attack one of my paralysing attacks would come on."

FIG. 3.—Facies. Case 3.

The patient's ailment has never prevented him from continuing at his work, and he does not complain of anything else than the symptoms described above. For years he has given up all thought of amelioration, having consulted many doctors and visited various hospitals without avail.

Previous history.—Rheumatic fever at the age of 9. Extremely moderate in alcohol. Venereal disease denied. No accidents.

Family history.—One of his five children died with rheumatic fever. Otherwise, negative.

Condition on examination.—The nervous system is normal in all respects. Optic discs normal. Ocular movements normal. Reflexes all normal. Has

not put on weight to any extent (see fig. 3). X-ray of sellar region shows it to be normal. Wassermann test in the serum, negative. Heart normal. Blood-pressure, 135 mm. Blood-sugar normal, and complete urin-analysis also (Dr. Lawrence).

This third personal case similarly conforms to the group of combined narcoleptic and tonelessness attacks, and is of especial value, in that it furnishes evidence of the occurrence of trance-like states independent of emotional excitation. It also is of peculiar interest owing to the transition from sleep attacks to "paralysing" attacks if the former are forcibly checked. When Cases 2 and 3 are compared it will be realized at once that between them they supply clinical data which throw light on the possible interrelation of the sleep and the emotional syndrome. To these matters we shall return immediately.

Case 4.—*Male case. Duration, two years. Cause, indefinite illness two or three months before, diagnosed as epidemic encephalitis. Typical short diurnal attacks of sleep, with attacks also of loss of power in the limbs (often local) under the influence of emotional stimuli.*

A. B. C. D., aged 18. Schoolboy.

History of present illness.—Nearly two and a half years ago the patient, when at school, suffered from some kind of febrile ailment, thought at first to be influenza but later asserted to have been epidemic encephalitis. No diplopia was complained of, nor was abnormal sleepiness or sleeplessness a symptom. Within a period of not more than a month or two, however, he put on no less than a stone and a half in weight, and at the same time began to have sudden attacks of "sleep" at meals, or when unoccupied or uninterested. An imperious desire to close the eyes was the aura, and immediately they closed he fell asleep. The duration was usually extremely brief, amounting merely to half a minute or even less, but on occasion it extended to a quarter of an hour. They have continued without intermission ever since, recurring several, sometimes many, times a day. In these attacks the patient is not as a fact deeply asleep: on the contrary, he is immediately aware when he is touched, when he is spoken to, when anything goes on round him; on a number of occasions he declares he has not been asleep at all, but has presented merely the appearance of sleep, for his state he thinks is more akin to that of "day-dreaming," a sort of "reverie" which is different from ordinary sleep. Once in a railway train the desire to close the eyes overmastered him, and instantaneously a book he was holding fell heavily from his hands; the abruptness of the onset of the sleep attacks always strikes him as peculiar. Night sleep is normal and of normal duration.

From the commencement of the affection another, to him very curious, symptom has manifested itself. It showed itself first on the occasion of a

school steeplechase. He had to jump a hedge, and as he approached it running he suddenly felt a momentary anxiety whether he could do it or not. On the instant his limbs relaxed and he fell helpless against the hedge, completely unable to get his legs to answer his call for an effort. Within half a minute he had recovered and surmounted the obstacle without difficulty. On many occasions since, a similar but very brief failure of power has made its appearance, notably when he is playing tennis. Just as he is about to serve the ball a thought flashes through the mind that he cannot do it and an equally momentary weakness of the right arm develops; by an effort he overcomes the loss of power and delivers his service usually so well that the momentary hesitation is scarcely observed.

The toneless or powerless attacks are thus largely local and of extreme brevity, but it is otherwise with the act of laughter. He has cultivated what he calls an "artificial" or "unnatural" laugh, because on one occasion when laughing heartily he became alarmed at a sudden weakness in both his legs, so that he nearly fell to the ground. As there were onlookers he felt highly sensitive about it; he now avoids the possibility of a recurrence by laughing in an unnatural way, "outwardly" and not "inwardly." He has some difficulty in describing exactly what he does at such times, but it is sufficient to prevent the development of the phenomenon of temporary weakness.

Previous history.—With the exception of the illness already alluded to the previous history is negative, and there is no record of head injury.

Family history.—On both sides of the family cases of "bad temper" in an aggravated form have occurred. His mother is an "exceptionally nervous woman."

Condition on examination.—A bright and highly intelligent boy, with a fine scholastic record, he shows no sign of any organic affection of the nervous system. All the usual methods of examination have proved consistently negative.

The clinical interest of Case 4 resides mainly in the inapplicability of the term sleep to the first kind of seizures, which more closely resemble reveries (though outwardly they are like the sleep state); in their absolute suddenness and brief duration; and further, in the almost larval or embryonic type of the cataplectic attacks, which are often scarcely more than momentary and may well pass unobserved; in their frequent restriction of range, and in the apparent success with which the patient manages to prevent them from reaching fuller development.

Classification.

In the present state of knowledge a complete ætiological or pathological classification is out of the question. We are compelled to fall back on an empirical clinical grouping. In my opinion a distinction into "idiopathic" and "symptomatic" varieties is not

calculated to further knowledge, for the very good reason that idiopathic narcolepsy cannot be causeless, and examples analogous to it in every respect have been seen to follow an attack of epidemic encephalitis. All narcolepsy is as a fact symptomatic, but at present we do not always know of what pathological or physiological condition it is thus symptomatic. In this, as in every scientific classification, the six-century-old aphorism known as Ockham's razor ought never to be allowed to slip from the mind: "Entia non sunt multiplicanda praeter necessitatem."

A purely tentative division along the following lines, however artificial, may prove clinically convenient.

(1) Recurring diurnal attacks of sleep: (*a*) associated with attacks of "tonelessness"; (*b*) without "toneless" attacks.

(2) Sleep attacks of prolonged duration, day and night sleeps running into each other; continuous sleep.

For various clinical examples of these hypersomnic conditions a pathological basis has been found, and this aspect of our subject will be examined in due course, but it has not yet been discovered for all. Further, the range of the pathological substratum is so extensive, covering cerebral tumours on the one hand, and psychopathological states on the other, that a classification founded on it would not provide any clearer idea of the essence of the disorders.

Taking the above empirical grouping, we may consider it in detail.

Clinical Varieties.

I.—Cases which definitely belong to the first subdivision of the first class (sleep attacks with attacks of "tonelessness") must still be regarded as relatively and absolutely rare. In his monograph Adie has given a précis of those (in males) reported by Gélineau [28], Henneberg [36], Goldflam [30], Redlich [70], Jolly [41], K. Mendel [54], Noack [62], Somer (2) [79], Loewenfeld [47], Stiefler (1919) [85], Stöcker [88] (his Case 4), and Singer [77], and has added one male case of his own—a total of 14, to which he appends three personal female cases—17 in all. Typical cases (in males) published before his own communication, but not included by him, are those of Curschmann and Prange [15] (their Case 2), Westphal [93], Carl Camp [9], Redlich (2) [69], Wenderowic [92], Matzdorff [52], and Bruno Fischer [23]—8 more; since his paper appeared there have been added those of Janzen [40] (his Case 2), Hilpert [37], Spiller (3) [82], and the first three here supplied by myself—a total of 30 male cases. In respect of this first

variety of narcolepsy in the female sex, Adie says that "hitherto the only published female cases were two described by Nevermann and by Kollewijn of "narcolepsy in pregnancy," but while these are judged by him not to have been cases of "true narcolepsy," "for sleep attacks alone occurred and they ceased after delivery," he has overlooked those recorded by Kahler [42], and by Guleke [35], both of which are highly characteristic, as well as the female cases of Gowers [33], and others (see below). Recently a typical case in a woman who subsequently developed disseminated sclerosis has been briefly recorded by Jacobsohn [39]. This gives us 6 cases in the female sex, to set against 30 male, a proportion of exactly 5 to 1 in favour of the latter.

There fall also to be included in this same clinical group those which have seemed to be consecutive to an attack of epidemic encephalitis, viz., the cases of Stiefler [86] (1924), Missriegler [55] (this case, of much interest, has been seemingly overlooked by both German and English workers), Adie, Münzer [59], Symonds [89], Stiefler [87] (1926) (his Case 2) and my fourth case, seven in all, and all males. This raises the sex ratio to over 6 to 1.

In every one of these forty-three male and female cases (I do not doubt that I may have overlooked some others unintentionally), the clinical record specifically states that in addition to recurrent diurnal attacks of paroxysmal sleep, the individual concerned exhibited in greater or less degree the phenomenon of loss of tone in the skeletal muscles (in one or other part), under the influence of stimuli of the emotional series.

II.—Taking next the second subdivision of the first class (and here I shall include both those cases in which the "toneless" attack has been looked for but not found, and those in which no reference is made to it), we find numerous instances in both old and recent literature. The recent cases are those of Adie [1], Redlich (his Case 3) [69], Goldflam (2) [30], Curschmann and Prange (their Case 1) [15], Janzen (his Cases 1 and 3) [40]; and, among the older ones, after having read through all the original records, I shall include those reported by Chavigny [13], Foot [25], Morton [58], Caton [12], Ewen [21], Pitres and Brandeis [66], and Rybakoff [74]—all in males. The female cases of this subdivision comprise those of Gowers [33], Blodgett [4], MacCormac [50], and possibly that of F. Fischer [24]. In all of these, male and female, no cause for the narcoleptic manifestations was discoverable. But when we come to similar cases in which an association with a definite pathological state seems established,

the numbers increase materially. Of the postencephalitic cases we may enumerate those of Stiefler [87] (his Case 1), Perrier (2) [64], Wenderowic [92] (his Case 2), and Redlich (this case appears to me uncertain); typical cases developing on a basis of commotio cerebralis have been published by Lhermitte [46]; others as clearly linked to one or other variety of epilepsy are those of Féré [22], Gowers [32], Worster-Drought [96], Goldflam [30] (his Case 5), Brailovsky [5] (some others here are omitted as being rather indefinite); cases arising on a hysterical or other psychopathological basis have been reported by Camuset [10], Ballet [2], Laehr [43], McCarthy [49], Nammack [61], Carlill [11], Myers [60], Worster-Drought [96], Brailovsky [5], and many more; a syphilitic infection was thought to be responsible in one of Lamacq's [44] cases. It would be easy, indeed, to enlarge still further this class of recurrent diurnal narcolepsy of comparatively brief duration, for I have made no reference to arteriopathic hypersomnia, or to the sleep attacks of certain forms of drug-taking, or of certain cases of cerebral tumour. Most of the cases of the latter two categories, however, come in reality under our second clinical division. What impresses the student when he surveys this large class is, that as far as recurring attacks of irresistible diurnal sleep are concerned, there is no affinity whatever between them and any one, specific, invariable form of disease.

Seeing that brief attacks of diurnal sleep can and do occur as a monosymptomatic variety of narcolepsy, it becomes a question of considerable interest whether the *Tonusverlust* that develops on the heels of strong emotion may not also be found monosymptomatically; if so, what place is it to be assigned in the narcoleptic category? As far as I can ascertain from a complete survey of the literature, no case of attacks of loss of power or tone through the influence of emotion, occurring by themselves, is on record.[1] But attention may be directed to the following clinical data:—

(1) In Somer's second case, that of a youth of 20, the "toneless" seizures took place not only under the influence of emotion but also after the patient had been walking or standing for a while, being then unaccompanied by any obvious affective experience; further, they were noticeably more prominent in the picture than the attacks of sleep, which by comparison were few and far between.

[1] Since this paper was written a case of cataplexy has been published by R. D. Gillespie (*Journ. of Neurol. and Psychopath.*, July, 1927) in which "affective attacks" occurred practically alone. Yet sleep apparently of abnormal quality was an occasional afternoon occurrence.

(2) In Symonds' post-encephalitic case the two types of attack occurred at different periods, for when the patient came under observation he was suffering only from the affective seizures, the sleep seizures having ceased and being replaced by insomnia. Conversely, the illness in Hilpert's case commenced with the development of affective attacks, which eventually disappeared, although the sleep attacks continued.

(3) Berliner, Hoff and Schilder [3] have recently described a case of monosymptomatic *Tonusverlust*, which they reckon "in die nächste Nahe zum Tonusverlust des Einschlafens und damit zum Tonusverlust der Narkolepsie." The attacks in this case (an organic one) appear to present an intimate similarity to those of the narcolepsy group, but sleep attacks did not occur.

Clearly the effort to circumscribe the conception of narcolepsy alluded to above becomes more dubious than ever if cataplectic seizures can appear by themselves, or be a feature of other states than narcolepsy proper. Further, in this same connection, the importance of *larval or incomplete varieties of narcolepsy* must be underlined. The following personal case, recently observed, seems to me particularly illuminating.

Case 5.—Male case: larval or incomplete variety. Duration, three years. Cause unknown.

H. G. B., aged 32. Occupation, business.

History of present illness.—For about three years the patient has noticed the occurrence of peculiar attacks, of brief duration, during which he seems to be unable to move his eyes; he finds himself staring at some object, and in a kind of day-dream; the object then becomes indistinct, as if seen through a haze, and only by a strenuous effort can he cut short this state and break into its fixity. If no effort is made the phenomenon disappears spontaneously within a minute or two. He has also observed that on occasion this immobility seems to spread so as to involve other musculatures, giving him the feeling that he is quite unable to move any part of his body, yet at the same time the thought comes into his mind, "Oh yes, I can move if I like"; as a fact, however, he always remains motionless for an appreciable period, measurable in seconds. No emotion of any conscious kind initiates these spells.

For about the same time (three years) he has repeatedly noticed that under the influence of any excitement or other emotional stimulus he turns deathly pale and loses control over his limbs; in his own graphic language, "I feel as if I had no knees"; when playing a game at bridge, wondering what sort of a hand he is going to pick up, he feels his legs "disappearing" and is thankful he is sitting on a chair. With some five or six friends he meets regularly at a

restaurant for coffee, and the decision as to who pays is reached by one of the circle piercing the menu card with a pin haphazardly; the letter nearest the pinhole is taken, and the letter of the respective surnames of the members nearest that in alphabetical order decides. During this trifling procedure he has frequently turned "as pale as a sheet" and, standing on one occasion, had to hold on to the furniture as his lower limbs were sinking. His friends have noticed his demeanour and uncharitably attributed it to unwillingness to pay, whereas in fact it has been the outcome of the affective experience. Hearty laughter produces the same result.

On no occasion, as far as can be ascertained, has the patient had sleep attacks; yet many times during the day he suddenly becomes conscious of "a warm feeling round his eyes," leading him to close them for a moment, and he remarked spontaneously that he always feels that if he chose to he could then allow himself to sleep. As a fact, he has never thus succumbed to the impulse.

Family and previous history.—Negative.

Condition on examination.—A normal finding in all respects.

Had our knowledge of narcoleptic syndromes not widened in recent years I doubt whether the real nature of this case would not have escaped me; as it is, I confidently assign it to an abortive or incomplete class in the general group, at the same time stressing the point that *only the affective or "Tonusverlust" attacks have passed into a characteristic stage.* And it is a further matter of no little interest that this patient also exhibits almost a cataleptic immobility (cf. my Case 3 above) which, brief or momentary as it is, is apparently independent of emotional excitation.

III.—If we take, next, sleep seizures of a prolonged or continuous kind, we are familiar with their occurrence under equally variable pathological circumstances. Clinical distinction here between natural sleep and states of stupor (anergic, catatonic) or of semi-coma is often enough difficult to establish, and the same is true of cataleptic or trance-like conditions and of those associated with drug addiction. Prolonged hysterical sleep is well known and has been the subject of numerous communications (e.g. Raecke [67]); in this connection no recent writer has mentioned the case of the "sleeper of Oknö," as narrated by Fröderström [27], who slept for thirty-two years. The curious will find in the volume by Gould and Pyle [31] valuable historical references to cases of protracted sleep of the hysterical, cataleptic, and stuporous kind. Similar more or less continuous sleep states also accompany some cases of such organic intracranial conditions as cerebral tumour or abscess; they have been known for many years, and have been made the subject of numerous useful studies (Righetti [72], Franceschi [26], Soca [78], Purves Stewart [83];

more recent communications are those of Bychowsky [8], Pette [65], Lucksch [48]; Thomas, Jumentié and Chausseblanche [90]; Souques, Baruk and Bertrand [81]).

Finally, I need here only mention the well-known short or, much more commonly, protracted sleeping states of epidemic encephalitis and of trypanosomiasis to complete the catalogue of the narcoleptic syndromes belonging to the second clinical group.

Clinical Symptoms.

(I) *Sleep.*

Although the term sleep is freely applied to the phenomena of the great majority of the cases usually classed as of narcoleptic character, the ostensible reasons for this require to be examined. It must be apparent to any critical mind that no definite clinical distinction can be drawn between degrees of sleep, which range from merely "dozing" with the eyes shut, on the one hand, to a profundity of unconsciousness not easily to be separated from a comatose state, on the other. In clinical practice distinctions and contrasts are far less easy to draw than they are on paper. Can sleep as a fact be readily differentiated from sleep-like states, and if not, where are the latter to be considered as beginning or ending? In the direction of deepening unconsciousness one passes by easy gradations from somnolence to sopor, sopor to stupor, and stupor to coma, as Monakow [57] long ago emphasized; but if the sleep of narcolepsy is thought to be distinguished by the fact that the patient is easily roused therefrom this is in the first place *not* true for every case, and in the second characterizes some other unconscious states not of the same nature, e.g., some forms of hysterical fit. Glancing through German literature one finds such terms as *Schläfrigkeit*, *Schlafsucht*, *Einschlafsucht*, *Somnolenz*, *schlafähnlicher Zustand*, *Schlafanfälle*, *Torpor*, *Lethargie*, *Sopor*, in frequent use, without their apparently corresponding to any clear-cut distinctions of a clinical character. If, again, we take the descriptions of cases confidently assigned to the group of narcolepsy it is at once evident that in quite a number of instances the attack of sleep does *not* correspond to ordinary conceptions of that state, for we note that frequently the patient is declared to "have heard what was being said," or "to have been aware of what was going on round about him" (Goldflam, Westphal, Redlich [68], Somer, Chavigny, Mendel [54], and others). Even a normally light sleeper is not in point of fact asleep if he is cognizant of

what is going on. It thus becomes more than doubtful whether the sleep of the narcoleptic is to be likened in all respects to normal, healthy sleep.

Equally remarkable, and somewhat disturbing to any view of narcolepsy as just an exaggeration of a normal cycle of sleeping, is the significant observation made by not a few that the patients declare they exhibit only the appearance of sleep: they are in reality aware of their surroundings *but not able to move a single muscle or articulate a single word* (Redlich [69], Guleke [35], Bruno Fischer [23], Loewenfeld [47], and others). That is to say, the condition bears only an apparent resemblance to sleep and turns out to be transitional to the cataplectic class of seizure. In my Case 2 the attack of tonelessness looked rather like sleep, inasmuch as the patient's eyes were closed and his head nodded on his breast, whereas as a fact he was entirely conscious and awake, and the physiological phenomena were of a peculiar kind. Again, in my Cases 3 and 5 trance-like states occurred, apart from emotional excitation, which bore the external marks of sleep, but their content was altogether different, seeing that in point of fact the patient was fully conscious of his surroundings but unable to move a muscle. The "sleeping" and the "toneless" attacks are evidently linked to each other, in many cases, much more intimately than appears from a casual survey. Thus this interesting condition, found in many cases of narcolepsy, widens the conception and approximates it to that of cataleptic and trance-like states, to which some erroneously think it is altogether foreign. This aspect of the problem is of considerable significance.

Another argument can reasonably be advanced against the hypothesis that the sleep of narcolepsy is ordinary sleep, pure and simple. The normal subject falls asleep because he is sleepy, whereas the great majority of narcoleptics are not conscious of undue sleepiness as a prelude to the attack of sleep; nor, for that matter, curtailed as the duration of the sleep nearly always is, are they sleepy or drowsy thereafter; on the contrary, most of them say they are at once quite fresh. And again, the vast majority enjoy sleep of normal duration at night, and never oversleep themselves. It is obvious, in short, that the abnormal slumbers of the day differ from nocturnal sleep in several respects, the possible significance of which it would be well not to underestimate. Suddenness of onset of the diurnal sleep has characterized very many cases; two of my patients have repeatedly fallen asleep at meals, with their food halfway to their mouth. A quite

forgotten case reported more than seventy years ago by Graves [34] is that of a gentleman who had sudden attacks of unconquerable sleep; at meals he broke one wine glass after another by falling asleep after the act of filling the glass and during the time he was raising it to his mouth—a literal slip 'twixt the cup and the lip. This abruptness of onset is highly characteristic. There are but few exceptions to the generalizations just mentioned. Persistent sleepiness is distinctly rare, as is physical tiredness. Stiefler's [85] patient (1918) would fall asleep after the Sunday midday meal and remain in unbroken sleep till the next day; he was nearly always tired and sleepy, but was definitely alcoholic, and there was a family history of alcoholism in father, mother, and three brothers. Caton's [12] patient had a constant sleepy look and could only keep himself awake by active exercise or mental stimuli. Tiredness preceded the attacks of sleep in the cases of K. Mendel, Goldflam, Jolly, and some others. In a few cases the conditions and circumstances tending to lead to sleepiness in normal persons (monotony, warmth, post-prandial repletion) have been noticed to affect narcoleptics also, but in numerous other instances no such association has been found (cf. as an instance my Case 2; the patient fell asleep as readily during a business conversation as when at the pictures: whether he was warm or cold made no difference). Bruno Fischer's patient sometimes opened his eyes during the sleep attack, so that his friends did not always know whether he was asleep or not.

Because of these peculiarities and differences we are compelled to question any strict identification of the phenomena of so-called narcolepsy with those of normal sleep in the healthy individual, close though the superficial analogy may be, and frequently is. Had we any objective criteria to fall back on, differentiation might be more readily accomplished, but the sign of contracted pupils in sleep, relied on to some extent by Goldflam because noted by him in his cases of narcolepsy, has not a universal applicability. If the useful researches of Richter [71] on changes in the electrical resistance of the body during sleep are corroborated, it may be that along these lines light will be thrown on differences between normal sleep and certain pathological conditions nearly resembling it. He has found, for example, a gross difference in the electrical resistance of the skin in a number of patients in catatonic stupors, "some of which so closely resembled sleep that it was impossible to tell by looking at the patients that they were not asleep," as compared with the resistance during normal slumber.

Enough has been said, however, to cast doubt on all simplistic

interpretations of the diverse sleep phenomena of narcolepsy—phenomena not embodied in the terminology—and to suggest we are dealing with syndromes not consisting in recurrences of natural sleep but presenting features which offer a link of union with other psycho-pathological conditions of particular interest.

(II) *Attacks of Loss of Tone and Power.*

This feature of our first variety of narcolepsy is of consummate interest, because, or in spite, of the fact that it bristles with difficulties.

In the first place, subjective descriptions apart, clinical observation reveals modalities and divergencies of considerable importance. Perhaps the commonest type of attack is that in which, under the influence of emotional excitement, a toneless and powerless state of the skeletal musculature supervenes, with the consequence that the person concerned finds his knees giving way and he sinks to the ground; after a brief period, measurable usually in seconds, tone and power return and he is able to get up again. Attacks corresponding to this general account have been remarked in all of the forty-three male and female cases which I have collected, but they may vary materially in degree and extent. Sometimes the patient is conscious of no more than a sinking at the knees, possibly scarcely noticeable to the onlooker; in other instances the seizure is complete, affecting neck, trunk, arms and legs and lasting for quite several minutes. In one and the same case the degree may differ at different times and under differing circumstances.

The actual clinical state is one of transient, combined diminution or loss of volitional power and of muscle tonus, and it is for practical purposes difficult to separate the two components. With complete outfall of tone some loss of power must ensue (cf. the apæsthesia experimentally produced by Mott and Sherrington), though the two need not proceed *pari passu*. In respect of the average condition in narcolepsy objective examination reveals both a loss of power, amounting to absolute immobility, and a loss of tone; the observation made by myself and described fully on a preceding page is of high significance, for it furnishes proof of tonelessness so profound as to result in temporary abolition of the deep reflexes. Numerous observations point to the occurrence of partial and segmental failure of power and tone. The incidents mentioned in my Cases 2, 3 and 4, are paralleled by analogous occurrences in the cases of Janzen (his Case 2), Wenderowic, Goldflam and others. In all of these power went out of the limb or limbs about to be innervated volitionally—a kind of motor inhibition, under the influence of excitement.

A second feature of the cataplectic attack is that in various recorded cases something more than, or different from, pure loss of tone takes place. Trembling (Westphal), movement of the head and protrusion of tongue (Noack), twitching movements of eyelids and face (Bruno Fischer), of lower jaw (Wenderowic), and other involuntary movements (Redlich [70], Carl Camp), have all been seen. The possibility, also, of *voluntary movements of correction* complicating the muscular weakness is specially mentioned by Adie. The absolute speechlessness of many cases is not encountered in all; my patient (Case 2) made a grunting noise as his head fell forward; Somer's patient muttered something unintelligible. An equally interesting observation is that of Redlich [69], who in one case noted that hearty laughing led to an attack of powerlessness which culminated in involuntary relaxation of both vesical and rectal sphincters.

One of the integral characteristics of the phenomenon is the full conservation of consciousness during its course, however much, in some cases, the appearance of the patient may belie this.

The exciting cause of the attacks is declared almost universally to reside in a stimulus of the emotional series, among which anger, annoyance, compassion, agitation of any kind, amusement or joy may be specially mentioned. The *act* of laughing provokes the attack more readily than any other cause, but we must note particularly that in this case the emotional stimulus itself, unless it manifests itself in laughter, does not suffice; in other words, while anger, and excitement generally, will precipitate the attack in spite of the patient's efforts to frustrate it, a stimulus arousing laughter will not produce the attack should he be able to inhibit the motor act of laughing. A consideration of the clinical data undoubtedly bears this contention out. In a number of cases it is expressly stated that only irresistible laughter culminates in the loss of power. Only when Gélineau's patient "riait aux éclats" did his limbs give way. My patient, E. C. (Case 2), smiles readily enough, and laughs softly, without experiencing the cataplectic phenomenon. Stiefler's [85] patient never developed the affective attack from laughing, but only with rage or annoyance.

Somatic accompaniments indicative of the emotional disturbance, whatever its modality, are not referred to in the literature to any extent —a curious point well worth passing notice. I can find only the briefest allusion to these. One of Redlich's [69] patients said that in the seizure he had a feeling of "losing his breath"; another said he was conscious of a "warmth spreading over him." In Wenderowic's

case the patient was aware of a "pulsating noise in his head": Somer's second patient showed pallor of the face, as does one of mine (Case 5). The case recorded by E. Mendel concerned a patient who exhibited attacks of *Tonusverlust*, during which there was pallor, the pulse was slow, and perspiration broke out at its close. My own patient, T. T., described with great clearness palpitation of the heart and a feeling of tingling over the head, cheeks and body. Doubtless somatic phenomena of these and similar kinds occur more frequently than is recorded, yet the speculation arises whether, in the case of cataplexy, the emotion does not manifest itself abnormally, to the relative exclusion of the usual outward accompaniments.

(III) *Transitions and Substitutions.*

Cursory consideration of the two types of attack might yield the impression that they consist of forms essentially dissimilar, correlated only through their occurrence in one and the same case. This is without doubt a fundamentally erroneous conception, as can at once be realized by attention to the following clinical variations.

(1) *Emotional excitement may lead not to the cataplectic but to the narcoleptic state.* This occurred with regularity in the cases reported by Kahler and Westphal. Under the influence of emotion the patient whose case is reported by Bruno Fischer felt a strong inclination to sleep. The connection is shown best of all in Gélineau's original case: "s'il a une émotion profonde, pénible ou joyeuse, le besoin de dormir est encore plus impérieux et soudain."

(2) *Sleep may be associated with or follow the cataplectic state.* During the attack of powerlessness following laughter, Matzdorff's patient felt an irresistible desire to sleep.

(3) *If sleep is prevented the cataplectic attack may ensue.* This interrelation is exhibited in perfection by my patient E. C. (Case 2 above). Similarly, in the case of T. T. (Case 3 above), interference with the impulse to sleep led to the oncoming of the "paralysing attack," to use the patient's own expression.

(4) *The cataplectic state may develop spontaneously, as does the narcoleptic attack in most cases, and be unaccompanied by emotion.* This occurred in the cases of Goldflam and of Somer (his Case 2); and in my Case 3, as we have seen, the "paralysing attack" bore no relation to emotion, as far as could be ascertained. A similar remark is applicable in respect of my Case 5.

(5) *Many, both of the sleep and the cataplectic, attacks are in reality*

identical in comprising a component which corresponds to the usual conception of catalepsy or trance. In a previous section I have furnished evidence fully bearing out this statement; awareness of surroundings, coupled with inability to move, has marked one or other type of attack in the cases of Nammack [61], Westphal [93], Somer [79], Guleke [35], Gélineau [28], K. Mendel [54], Goldflam [30], Redlich [69], Chavigny [13], Bruno Fischer [23], Loewenfeld [47], three of my five personal cases and others. The association occurs with an impressive frequency, and cannot be dismissed as accidental.

Consideration of these various divergencies must in my opinion compel us to envisage the possibility that a common factor may yet be found for all the clinical phenomena; that some not as yet clearly understood inhibitory mechanism is responsible both for the "sleeping" and for the "falling"; that the relations between narcolepsy and other clinical syndromes now to be examined are so close as to render strict delimitation of the former both impracticable and inadvisable.

Relation of Narcolepsy to other Clinical Syndromes.

(1) *Relation to myoplegia.*—As far as I have discovered, Wenderowic is the sole author who makes reference to the resemblance between the attacks of *Tonusverlust* and those of myoplegia paroxysmalis or periodic family paralysis. Yet the analogies are impressive, in spite of the considerable differences. The examination of the phenomena which occurred in the course of the cataplectic attack of my Case 2 at once suggested a certain similarity to those of myoplegia, since in their essentials they consisted of a sudden, widespread, flaccid "paralysis" of skeletal muscles with complete loss of the deep reflexes. The time was much too short for any investigation of the electrical reactions; some future observer may be able to furnish us with data in this respect. In the usual family myoplegia the attack is crescendo in character, taking, perhaps, hours to develop and hours to die away, but at its height the combination of flaccid palsy with loss of deep reflexes is identical with that reached in a few seconds in the case of cataplexy. I purposely do not emphasize these resemblances, the divergencies being obvious to everyone, yet it is well to make a note of them; according to Wenderowic a relative lymphocytosis characterizes myoplegia, and a similar blood-picture has been found in narcolepsy by Janzen and by Stiefler [87]. The significance of this blood-change is as yet indeterminate.

(2) *Relation to catalepsy.*—This term, like too many other medical

expressions, suggests a seeming precision which, as a fact, it does not possess. By catalepsy is usually understood a state of motionlessness, of sustained immobility, with or without clouding of the sensorium; the patient exhibits the appearance of sleep, is commonly indifferent to stimuli, and often shows the phenomenon of flexibilitas cerea in the limbs. As a symptomatic term the word has its value, indefinite though it be; the difficulty is that both catatonia and catalepsy convey the idea of the presence of the flexibilitas cerea just mentioned; yet this is not a cardinal, in the sense of exclusive, feature of the condition, as I have seen it in the sleep states of epidemic encephalitis, in frontal lesions of organic character, and in other intracranial states (cf. Brissaud [6]). We need an expression to signify the condition of being "struck all of a heap," of immobility, following on affective shock, which is sometimes met by use of the phrase "catatonia attonita" (cf. *Schreckkatalepsie*). Doubtless catalepsy covers a multitude of dissimilar syndromes, but its main connotation, as given above, has received general acceptance. Many catalepsies are hysterical; the majority, indeed, are, widely speaking, psychopathological. Now we have already seen how many of the attacks of the narcoleptic are signalized precisely by this trance-like state of awareness of environment coupled with absolute inability to move a finger. No allusion is made by any modern writer on narcolepsy to the once much-discussed question of *nocturnal paresis* or *paralysis*, which formed the subject of an interesting study by Weir Mitchell [56]. While doubtless here also a number of differing clinical conditions are comprised, among them is one to which I have long given attention. I have found it in various persons, healthy and otherwise, and do not regard it as anything else than a transient physiological disorder. It consists, briefly, in a state which supervenes commonly in the morning; the subject is awake, because he is conscious and knows where he is, but for the moment he is utterly incapable of innervating a single muscle. He lies in appearance still asleep, with eyes closed, yet within is engaged in an intense struggle for movement, with which is associated an acute mental anxiety; could he but manage to move a limb through a fraction of an inch, the spell would be instantaneously broken, and with a sigh of relief he would regain full power. I have had graphic descriptions of this "sleep paralysis" from more than one patient.

Its resemblance to the "paralysing attacks" of my third narcoleptic (T. T.) is of the closest. It is a physiological catalepsy, a state of sustained immobility with sensorial awareness, and as such is of much

interest for our subject. In a large number of the recorded cases the remark is made that the patient can be roused from his narcolepsy or his trance-like condition by a touch, a shake, a movement, whereas stimuli through other sense avenues are less effective. It seems to me this observation may have possible significance.

(3) *Relation to epilepsy.*—Much controversy has raged round the question of the relation, if any, of narcolepsy to epilepsy. On the whole, most authors deny any connection between the two. Curschmann and Prange go so far as to advocate abandonment of the term narcolepsy, because "it suggests a relationship to epilepsy which does not exist." Adie repeatedly stresses the point, declaring that it is highly important "we should distinguish it [narcolepsy] from a serious disease [epilepsy] with which it has no connection whatever." A similar attitude is assumed by Kahler, Goldflam and a number more.

But there is very much to be said against differential absolutism of this kind.

(1) The immediate and obvious answer to those who object to the idea of any association between the two is, what do they mean by "epilepsy"? Unless they furnish us with a definition of epilepsy it is useless to say that narcolepsy has nothing whatever to do with it. Yet in not one of the discussions on this matter have I found the objector to give a description of what he understands by "epilepsy": nay, further, I have noted frequently that epilepsy is still spoken of as a "disease," so little have Hughlings Jackson's doctrines made headway in this particular respect.

In my recent Harveian Lecture [94] on "The Epilepsies" (to which the reader is referred) I have advanced considerations, developed along lines suggested by Hughlings Jackson many years ago, which indicate the possibility of any motor cell-group becoming the seat of a "discharging lesion," so that an epileptic attack may take a number of seemingly very different clinical guises. The diseased condition (whatever it be) is the cause of the "epilepsy," which is a symptom pure and simple, and, as such, strictly physiological. Adopting this view of the question, I maintain strongly that *any alleged contrast between the narcolepsies and the epilepsies is merely a contrast between symptoms and leaves untouched the problem of whether or not there is a similar mechanism of production behind each.* That there may be, in point of fact, some such similarity of inhibition or release of function is surely conceivable and therefore worthy of discussion. This pathogenic aspect is dealt with later.

(2) The clinical observer must admit general resemblances between the two classes of symptom. As Hilpert [37] has recently remarked, apropos of narcolepsy: "Die Beziehungen zur Epilepsie sind demnach in jeder Hinsicht offensichtlich." It is indeed impossible to overlook the general analogies, which consist in periodical recurrence of attack, abrupt onset with or without aura, loss or diminution of consciousness, relatively brief duration; like many epileptics, many narcoleptics are fresh again soon after. The "fit" of sleep is in these respects like many "fits" of another kind. Of course the actual *content* of the "fit" is different in the two cases, but this does not affect the argument; for that matter, as I have said elsewhere, the word fit "might be legitimately employed to designate certain attacks of which motionlessness is the prominent feature." Besides, the involuntary movements of some at least of the attacks belonging to the cataplectic series offer more than just a casual resemblance to those of petit mal or of some mild epileptic seizure. Symonds observed one of these in his case, and took it to be epileptic, like petit mal; and I have already mentioned at the outset of the attack in my Case 2 the noise in the throat, the nodding forward of the head, the sinking down in a seemingly unconscious state —phenomena the analogy of which with some of those usually called epileptic it is pointless to try to controvert. If behind the ordinarily great dissimilarity of the external manifestations there is reason to postulate the action of not dissimilar mechanisms, it becomes unscientific to argue for fundamental differences between them from a nosological standpoint.

(3) Amplifying this last consideration, we may examine one or two of the arguments advanced by the supporters of the "no relation" theory. Adie says, "A most important feature of idiopathic epilepsy is that the attacks come on without apparent cause, whereas in narcolepsy the cataplectic attacks are 'reactive,' i.e., there is a definite exciting cause, namely, emotion." And again: "In genuine epilepsy the attacks are never reactive: this feature alone is sufficient to show that the two conditions are fundamentally distinct." But we have already noted that cataplectic attacks may develop spontaneously (Goldflam [30], Somer [79]), while the trance-like states of my Cases 3 and 5 also thus developed without any emotional antecedent. In the second place, the view that idiopathic epilepsy is never reactive is contradicted by familiar experience. We need not go so far as Rows and Bond [73], who explicitly affirm that "a survey of a large number of cases [of epilepsy] . . . . has demonstrated that some emotional

state involving a disturbance of consciousness, and some reaction to express the emotion, have been found in every instance." But of the occurrence of what we may call reactive or psychogenic epilepsy ("affect epilepsy") most observers have had abundant proof; it is recognized by all modern writers on the subject (notably Pierce Clark), and a good illustrative example was recently published in a paper of my own [95]. This particular criterion of differentiation, therefore, does not serve the purpose.

The claim that mental deterioration and psychial defects are characteristic of epilepsy but foreign to narcolepsy must be considered of relative and not absolute value, since numerous moderate epileptics suffer from their infirmity for years without exhibiting intellectual impairment. Much depends on the severity of the case, and on the actual class of attacks.

(4) More important, as forming a positive argument, is the fact that epilepsy and narcolepsy are on occasion definitely associated. For example, one of Gowers' [32] cases was that of a girl, aged 14, with a family history of epilepsy, who had suffered for four months from attacks clearly of that nature. But she had also, during the same time, "attacks of sudden pure sleep." When bromide was given regularly the epileptic seizures ceased, but the attacks of sleep went on unchanged. Failure of anti-epileptic remedies in narcoleptic cases cannot, of course, be advanced as an argument for the essential dissimilarity of the two conditions; some epileptics do not react to anti-epileptic drugs. Since the phenomena of narcolepsy are largely akinetic we should not expect sedatives to be the appropriate treatment. Besides, there is no greater ætiological fallacy than that *naturam morborum curationes ostendunt*. A valuable case is that recorded by Worster-Drought [96], in which the patient, a man of 25, suffered from fits in childhood up to the age of 10; since 1913 three fits ("description suggesting true epilepsy") have occurred, and also numerous narcoleptic attacks of variable duration, characterized by his falling asleep "at any time, under all conditions, and often in perilous situations." Once more, one of Goldflam's [30] patients (his Case 5) suffered from narcolepsy for six years, and later developed typical epilepsy with convulsions. Other instances of the same association are specified by Brailovsky [5] from records not accessible to me; the older cases of Féré [22] must also be alluded to in this connection, especially his Case 3, that of a youth of 21, who had both epileptic and narcoleptic seizures, the latter closely resembling normal sleep of brief duration (a quarter of an hour or less).

Féré specially noted that as the epileptic attacks increased the others diminished in frequency, and vice versa.[1]

During an ordinary major epileptic attack the subject is unconscious, while motor mechanisms escape or are stimulated into abnormal activity; during many narcoleptic attacks the subject is conscious, while the activity of lower motor mechanisms is in the completest abeyance. There is here, as I consider, a certain approach to the existence of "corresponding opposites," in Jacksonian phraseology, which may yet prove to be pregnant with meaning for an understanding of their physiological counterparts. Again, during the examination of my patient E. C. (Case 2), in the course of his affective attack I was able to demonstrate the temporary occurrence of an extensor response. This is known to characterize similarly many epileptic seizures, and it has, further, been observed in normal subjects during sleep. Collier [14] found in a number of healthy children up to the age of 12, during deep sleep, a typical "infantile response" (that is, extension); directly the child awoke the normal adult plantar reflex (flexion) occurred on stimulation. It cannot be without significance for our problem that a "physiological shift," as I call it, of the same type as is seen in normal sleep and in the epileptic fit, characterizes also the cataplectic seizure of the narcoleptic.

In face of these clinical observations it would be a mistake alike to ignore the similarities and to stress the differences; the correct attitude in my opinion is to suspend judgment and to await further contributions to our knowledge. So far, however, as the preceding considerations take us, they make out a *prima facie* case for the possibility of some of the narcolepsies and of the epilepsies having a common origin.

(4) *Relation to Oppenheim's Lachschlag.*—Under the term *Lachschlag* (*Lachschwindel, Lachohnmacht,* geloplegia) Oppenheim [63] described two cases (one of a girl and the other of a man, aged 45), of falling to the ground under the influence of hearty laughter. From the description furnished by the relatives (for no attack was personally observed) he supposed the patient was unconscious during the few seconds only of which its duration consisted, though he says in reference to the first case, "ob volle Bewüsstlosigkeit dabei eintritt, haben sie [the relatives] nicht festgestellt." Adie is inclined to look upon this *Lachschlag* as distinct from narcolepsy, whereas Jolly and Matzdorff both are impressed with the analogy it offers to the *Affektanfälle* of

[1] In a recent paper on "Epileptic Variants" (*Journ. of Neurol. and Psychopath.*, January, 1928, p. 231), I have recorded a case exemplifying a transition from epileptic to narcoleptic attacks.

the latter condition; Redlich [68] points out that the loss of consciousness in Oppenheim's cases was not absolutely certain, and mentions the cases in which sleep follows emotional excitation (see above); Goldflam, too, considers the resemblances of some significance.

I have myself observed one good case of the condition, that of a man aged 42, whose symptoms supervened on an injury to the head in the right frontal region. He noticed that when he laughed too loud he tumbled down, a condition which gradually so established itself that he had "dozens of tumbles." If he laughed slightly, he felt a sort of staggering feeling in the limbs; the louder he laughed the more he staggered; eventually he would fall to the ground, but would be on his feet again in a few seconds. Occasionally he "would go right off in the attacks," by which expression I understood from his description that he became unconscious of his surroundings for a moment or two, but not having seen the patient in such a condition I am unable to determine the exact state. By way of defence, he said to me, "I have cultivated a giggle to avoid the laugh." He had no attacks suggestive of sleep, and objective examination was negative.

The analogies which *Lachschlag* presents to the cataplectic attacks of the narcoleptic are much more impressive than the differences, so much so that it may fairly be considered a larval form of the other, or indeed, more or less identical.

Ætiology.

The search for possible causes of narcoleptic syndromes has revealed in some instances preceding factors to which the symptoms may legitimately be attributed, but in others none of recognizable pathogenic quality has been discovered.

As regards the former, *head injury* was noted in cases reported by Kahler [42], Redlich [69], and Singer [77], and occurred also in my Case 1 above. The appearance of narcoleptic phenomena after *commotio cerebralis* is mentioned by Souques [80], and has been made the subject of a careful study by Lhermitte [46]. The symptoms in the personal case of *Lachschlag* mentioned already followed an injury to the frontal region. *Alcoholism* in patient or parentage was a feature in those of Noack [62], Redlich [68], Stiefler [85], and Hilpert [37]. The possibility of an *endocrine* ætiology is suggested mainly by the interesting fact that in a considerable number of instances narcolepsy has been associated with a definite *increase in weight*, the result of the patient's becoming more fat (Hirsch [38], Ballet [2], Münzer [59], Caton [12],

Jolly [41], Stiefler [85], Lamacq [44], Redlich [69], Hilpert [37], Janzen [40], Curschmann and Prange [15], and my Case 2). The sella turcica was stated to be on the small side in those of Kahler [42], Redlich [69], Jolly [41], Singer [77] and Janzen [40]. In certain female cases the symptoms were aggravated during the catamenial flow (Fischer [24], Kahler [42]). A somewhat doubtful case reported by Dercum [17] was associated with an acromegaloid condition. We should mention, too, that a relative lymphocytosis of the blood occurred in cases published by Kahler [42], Stiefler [87], Janzen [40], and Redlich [69].

Attention must be directed also to the peculiar connection of some narcoleptic cases with *epistaxis*.

(1) The patient of K. Mendel [54] suffered from almost daily nose-bleeding between the ages of 8 and 22. When this finally ceased narcolepsy supervened.

(2) In a female case (MacNamara [51]) a girl, aged 15, had sudden attacks of profound sleep, which disappeared after a severe epistaxis, and did not return.

(3) Hilpert's [37] patient exhibited the typical combination of narcolepsy and cataplexy. An illness (? "cerebral influenza") developed intercurrently, associated with strong epistaxis, and thereafter the cataplectic attacks ceased altogether, though the narcoleptic continued.

(4) In consultation with Dr. Gordon Holmes I have recently seen a case of narcolepsy with cataplexy in a young man, with high blood pressure and slow pulse, who used formerly to have bouts of epistaxis regularly about once a month. Since the onset of the narcolepsy the epistaxis has entirely stopped.

These facts are rather striking, even though their significance at present eludes us.

Again, an origin for the narcoleptic syndrome in some disturbance of a *psychopathological*[1] kind was definitely ascertained in cases reported by Carlill [11], Myers [60], Worster-Drought [96] and Missriegler [55], and was probable in those of B. Fischer [23], Morton [58], Stocker [88] (his Case 4), and Brailovsky [5]. This aspect of the problem has undoubtedly been somewhat ignored by neurologists preoccupied with organic disease, and it might be developed further with advantage. I do not question that the so-called idiopathic

[1] Gillespie says of his case of cataplexy alluded to in the footnote on page 80, "psychological factors were evident enough, but psychotherapy had no lasting effect."

variety might be further dismembered were an adequate psychological investigation undertaken more frequently.

The data which have just been examined, together with what has been said on a previous page, apropos of clinical varieties, are sufficient to show how diverse and heterogeneous is the ætiology of the narcoleptic syndrome. To assort the causal factors is a matter of no little difficulty; a provisional arrangement might be made along the following lines:—

(1) Traumatic group.
(2) Psychopathological group
(3) Endocrine group.
(4) Epileptic group.
(5) Toxic-infective group (epidemic encephalitis, &c.).
(6) Circulatory group.
(7) Pressure group (cerebral tumours, &c.).
(8) Group in which no *Grundkrankheit* has been discovered.

Doubtless a cross-division may occur here; again, some ætiological components are surmised rather than proved in particular cases. In others the relation of the syndrome to the apparent cause, or to an outstanding feature of the individual case with which it may be linked, remains obscure, e.g., in respect of epistaxis and diplopia. I have already alluded to the former; of the latter it may be said that it was noted in cases recorded by Janzen, Spiller, Hilpert, and myself (Case 1). Whether it is of ætiological significance is undetermined; according to Janzen it is no more than a "prehypnotic" or "hypnagogic" diplopia, occurring in many normal people if they try to resist sleep. This explanation, however, does not appear to apply to my Case 1, in which the double vision was complained of independently of the sleep attacks, yet so far I have failed to discover any objective basis for it.

For the present the student of narcoleptic symptomatology must be content to recognize the diversity of the bases on which it arises in many instances, and to acknowledge his entire ignorance of them in others.

Pathology.

No autopsy on a case of narcolepsy belonging to the first clinical division has up to the present been conducted, so far as I know. Patients do not die of narcolepsy, which, as we have seen, is no disease.

What pathological information is at our disposal is derived from cases belonging to the second division, viz., cases of prolonged or

continuous sleep. The light they throw on those of the first group is oblique and indirect, and by itself but fitfully illuminates an obscure subject.

In 1900 a case of prolonged sleep associated with a basal cerebral tumour was published by Soca [78]. The patient was a girl of 18; during seven months "la malade dormait toujours, toujours de son sommeil paisible, d'aspect parfaitement naturel, toujours se reveillant facilement aux provocations ordinaires." The tumour was subsequently found to have penetrated into the third ventricle, and to have destroyed completely corpora mamillaria, tuber cinereum, chiasma and optic nerves.

Righetti's [72] case (1903) was that of a boy of 12, in whose case similar prolonged sleep was by far the most prominent symptom; "fino dell' ammissione il paziente e in uno stato di sonno continuo. La sue fisonomia appare calma, il respiro e regolare come quello di una persona sana che dorma." When called in a loud voice he would wake up, sigh, and fall asleep again. In this instance the tumour completely destroyed optic nerves, chiasma, bandelette, and tuber cinereum, while the walls of the third ventricle were distended. Basing his researches to some extent on statistics furnished by Schuster, Righetti collected 775 cases of verified cerebral tumour, of which 115 were noteworthy clinically for the occurrence of pathological sleep. He compiled the following figures, which I have embodied in a table.

TABLE I.

Site of tumour	Percentage of cases showing abnormal sleep
Thalamus and third ventricle	34·7
Medullary	27·9
Hypophysis and vicinity	26·5
Corpora quadrigemina and epiphysis	25·6
Cerebellum	15·8
Central gyri	10
Frontal	6, &c.

From these figures it is at once obvious that tumours situated in relation to the third ventricle (walls, floor, anterior and posterior extremities) together form a far higher percentage of abnormal sleep cases than any other group, and the association must be of significance.

Purves Stewart [83] published four cases of "tumour in the hypophysis cerebri" in 1909, and of these three were characterized by the occurrence of intense drowsiness, sometimes paroxysmal, lasting for hours, days, or weeks at a time. "In Case 1, and less strikingly in Case 4, the drowsiness had curious intermissions, during which the patient woke up for a time and was apparently clear and alert."

From the pathological descriptions the reader will find that while the pituitary itself was not invaded in all, the one common site of involvement was precisely the floor of the third ventricle.

Both before, and since, analogous cases have been put on record; present-day German writers on the subject confine their quotations largely to the recent cases of Pette [65], and Lucksch [48], which are not as a fact by any means so striking as some of those of which a précis is here furnished. Weisenburg [91] collected thirty cases of "tumours of the third ventricle" and found that "a decided tendency to drowsiness or somnolence" was specifically noted in sixteen, most of these being situated in the floor.

To correlate clinical and pathological findings in cases of cerebral tumour must be undertaken circumspectly; the effects of pressure, of œdema, of toxicosis, cannot be ignored, on the one hand, and, on the other, intracranial growths are commonly in the event too large for conclusions of localizing worth to be drawn therefrom. Thus it is that a tumour away from the third ventricle may be associated with somnolence or hypersomnia; in a case recorded by Bychowski [8] it was situated on the under surface of the left frontal lobe. Every neurologist can from his own experience quote cases of cerebral tumour of this kind; I may cite a single case of my own, in which the clinical symptoms closely resembled those of epidemic encephalitis as far as sleepiness and attacks of sleep were concerned, while the tumour proved to be located in the middle of the corpus callosum. Nevertheless the general conclusion holds, that basal tumours behind the chiasma are more likely than others to manifest themselves, *inter alia*, by hypersomnia.

It is equally hazardous to attempt to draw deductions from cases of cerebral abscess or of cerebral arteriopathy, for reasons which must be apparent. The evidence derived from epidemic encephalitis has been marshalled more particularly by Economo [20], whose general conclusion is to the effect that lesions in the vicinity of the third ventricle (floor, periventricular grey matter, region of the iter Sylvii) are responsible for the disorders of sleep that are highly characteristic of the average case of that disease.

Sleep Centres and their Localization.

Pathology can contribute comparatively little *specific* evidence, as we have just seen, but there are collateral lines worth scrutinizing. Of these one that appears to have some importance is the experimental,

though the data are rather meagre even if significant, and also now somewhat old.

Perhaps the most valuable work is that of R. Dubois [18], whose results were published as long ago as 1896, and have been neglected by all recent writers on the subject. Dubois experimented on marmots, and proved that "vers la partie antérieure de l'aqueduc de Sylvius et du côté du plancher du troisième ventricule, il existe des centres respiratoires de ralentissement et d'accélération, d'où dépendent également l'hypothermie et le réchauffement, la torpeur et la veille." He regarded the "centre" as a "centre du réveil," which plays "un rôle prépondérant dans le mécanisme du sommeil. Seulement son action est indirecte; il n'agit pas par une sorte d'inhibition imprimée par lui au système nerveux ou par une excitation centrifuge plus ou moins generalisée." This "centre" regulates respiration and circulation and produces natural sleep by a process of "autonarcose carbonique." Reviewing his experiments in a later publication, the same author states [19] that "J'ai mis hors de doute par l'expérimentation sur les marmottes un centre de sommeil, qui est d'ailleurs le même que celui du réveil, et se trouve justement situé dans la region où les auteurs que je viens de citer ont observé des lésions pathologiques coïncidant avec un sommeil persistant et prolongé." To Dubois without question must be assigned priority for some of the evidence now attributed to more recent work on the third ventricle and its grey matter.

Experiments conducted by Cushing and Goetsch [16] on hibernating wood-chucks dealt mainly with the pituitary gland, and led these workers to attribute hibernation to "a seasonal physiological wave of pluriglandular activity." It seems possible from the description of their operative technique that they injured also the tuber cinereum and other basal ventricular ganglia. Be this as it may, they found histological changes, especially in the pars anterior of the hypophysis in hibernating animals, as had Gemelli [29] some years previously in marmots. The latter, however, did not ascribe the somnolence of his animals to the changes in the gland, which he did not regard as a hypothetical centre for physiological sleep. Advances in knowledge of the last few years render implication of the pituitary problematical for these sleep states, and seem to invalidate the speculations of those who, like Salmon [75], attributed hypersomnia to functional insufficiency of that body.

Evidence has gradually been collected in support of the general

view already mentioned, a view to which a consideration of certain tumour cases, encephalitis cases, and Dubois' experimental work leads, viz., that physiological centres related to the floor of the third ventricle and the region of the iter—centres in the hypothalamus—have somehow a connection with the process of sleep. Undoubtedly much of this localization remains purely hypothetical, though it may well be of heuristic value. In the midst of conjectures (for pathological proof is wanting) it is desirable that the researches of Stief [84] should be given proper weight ; that observer has found constant, marked changes in the nucleus paraventricularis, and in the hypothalamus generally, in cases of general paralysis, senility, cerebral arteriosclerosis, various kinds of psychosis, cachectic states, &c., and while he believes they explain the development of vegetative symptoms, and possibly the onset of the senium, there is no mention of sleep as forming a feature of any of his cases. For the present, therefore, exact pathological knowledge does not take us very far along the road to a solution of the problem of what mechanism or mechanisms are responsible for the symptoms of narcoleptic patients, or of where precisely they are situated.

One of the major difficulties yet to be surmounted is constituted by our ignorance of the physiological processes underlying natural sleep. It is well not to ignore the extreme complexity of the factors, a point which of itself renders highly improbable the existence of a *solitary* " sleep regulation centre " localized with mathematical exactness towards the posterior-inferior end of the third ventricle. Sleep must be regarded as a complex physiological process, which can be influenced both by physicogenic and psychogenic excitations. Its actual phenomena comprise manifestations belonging to different series—respiratory, circulatory, muscular, sensory and psychical. To suppose all these varying phenomena to be somehow controllable by a relatively limited ganglionic centre puts rather a strain on physiological belief, even if some lines of evidence appear to indicate the possibility of a ventricular nucleus playing a definite though undetermined rôle in the *initiation* of the whole somatic process. We do not know whether this " centre " inhibits or releases, or what precisely are the mechanisms it controls. In view of these facts some of the contentions seriously advanced in recent communications will not bear investigation. Hirsch [38], for example, because his case of pathological sleep was found to be associated with an abscess of the left optic thalamus, says that " sowohl der mediale Teil des linken Thalamus opticus wie das in

der Nähe der Okulomotoriuskerne gelegene Höhlengrau Schlafzentren im richtigen Sinne sind"! "Localization" of this crude kind requires no comment.

We may, for the moment, consider Pavlov's views (1927), based on his work on dogs in connection with conditioned reflexes, and see how far they take us towards an explanation of the phenomena of narcolepsy. According to him, "sleep is nothing but internal inhibition which has become diffused continuously over the entire cortex and has descended also to some of the lower parts of the brain." Thus he considers sleep to be of *cortical* origin, but in its usual form simultaneous involvement of infracortical centres takes place, as evidenced by "relaxation of the skeletal muscles (closure of the eyes, drooping of the head, sagging limbs, and body limply hanging in the loops of the stand)." If, however, "a complete inhibition [is] confined exclusively to the cortex, without a concurrent descent of the inhibition into the centres regulating equilibrium and maintenance of posture (centres of Magnus and de Kleijn)," then a different form of "sleep" is observed; the animal "preserves an entirely alert posture . . . with wide open immovable eyes . . . remaining motionless sometimes for minutes and sometimes for hours. On changing the position of an extremity such extremity retains the new position." In other words, "the animal is in a state of catalepsy."

Interesting as these observations are, to what extent do they illuminate the problems of narcolepsy?

(1) As we have repeatedly observed, the "sleep" of the narcoleptic certainly in numerous instances does not conform to ordinary sleep, and in such cases there can be no universal cortical inhibition, with descent to centres regulating equilibrium and posture. Various examples of the narcoleptic keeping on his feet, nay, walking along, when "asleep," might be cited. To hold that in such cases the internal inhibition is confined to the cortex, subcortical centres remaining in activity, is illegitimate, for as we have just seen this physiological condition results, according to Pavlov, in catalepsy, which is signalized by absence of movement.

(2) Pavlov's experiments do not help us to understand the phenomena of cataplexy, unless by analogy we are to allow that internal inhibition might invade the "centres of Magnus and de Kleijn" while the cortex is as a fact functioning normally, in the sense that the patient is conscious and in a state of awareness. If so, there is here no question of "descent" from cortex to mid-brain, and the speculation is physiologically unsupported, as far as Pavlov's experiments go.

On the other hand, I attach importance to his physiological explanation of catalepsy, which condition I have tried to show is unquestionably linked to narcolepsy. The passing attacks of immobility, trance-like states, distinguishing more than one of my own cases and noticed also in others, receive here a possible elucidation.

Pathogenesis.

At this point it is well, I think, to recapitulate from the clinical aspect the salient features of narcoleptic cases, lest any preconception of what is implied in the term "sleep" should mislead us.

(1) In the great majority of instances the *nocturnal* sleep of the narcoleptic is normal. We must, therefore, consider that we are dealing with mechanisms that are disordered only during the day, and not at night. This fact of itself disposes of the contention that in our first clinical division of narcoleptic cases any structural impairment, or any more or less permanent abnormal physiological state of the mechanisms concerned, is in existence.

(2) "Sleep" is a complete misnomer for the attacks in a large number of cases. In many, the patient is not asleep, but motionless and aware of his surroundings.

(3) A whole clinical group is constituted by cases in which attacks both of sleep and of tonelessness occur. Not only are these in reality less separable one from the other than appears on superficial consideration, but emotional excitations sometimes cause one to develop, sometimes the other. Again, the toneless attacks may supervene without obvious affective antecedent, as may the sleep; on occasion, one may be exchanged for the other.

(4) All gradations are met with, from narcolepsy without cataplexy, to cataplexy without narcolepsy.

(5) An outstanding clinical feature of both types of seizures is their relative brevity, their diurnal periodicity, and the rapidity with which the patient recovers from them.

With these considerations in mind, we may in conclusion attempt an approach to the problem of pathogenesis.

I am in accord with previous observers who have commented on the fact that affective hypotonia is seen from time to time in normal people, or who have referred to the embodiment in colloquial terminology of the existence of emotional influence on body musculature. Affective *Tonusverlust* certainly occurs unspecifically in many ways. From this viewpoint, its occurrence as a feature of narcoleptic syndromes

is merely an exaggeration of the normal, and forms a difference in degree and frequency but not in kind. Laughing, tickling, fright, fear, may in the normal person lead to a helpless, toneless, motionless state of skeletal musculature. German writers refer to the *Totstellreflex* of the frightened animal, "frightened out of its life," as we say. Allusion was made above to the occurrence of sphincter relaxation during the cataplexy in one of Redlich's cases; this also can scarcely be judged pathological if we recall certain slang expressions that seem to be common to many races ("auch besteht im Hollandischen ein Volksausdruck welcher sich auf die Inkontinenz beim Lachen bezieht."—Janzen). A patient under the care of Brissaud [7] had a similar experience.

"Une dame lui annonce que son petit chien vient de mourir. 'Vraiment,' dit-il; 'oh! que c'est regrettable!' Est-ce le dernier mot 'regrettable' qui, par une sorte de suggestion, réveille en lui une série d'idées tristes? Toujours est-il qu'il se produit comme un déclenchement de tous les ressorts expressifs; la simple tristesse de la physiognomie entraîne les larmes; après les larmes, les sanglots; bien plus, ce malheureux homme n'est plus maître de ses sphincters. . . ."

Evidently, then, the affective or cataplectic attacks are not essentially pathological, and unlike anything seen in normal persons; on the contrary, the mechanism or mechanisms in activity at such times may on occasion be in action also under conditions that cannot be regarded as abnormal. In my opinion *this* is the main distinguishing feature between narcolepsy and ordinary epilepsy, considered as symptoms; the narcoleptic attack (either narcoleptic or cataplectic) reproduces a phenomenon to be seen also from time to time, in major or minor degree, in persons of normal neural function, whereas the epileptic attack does not. It is essentially abnormal, and not to be likened to any neural phenomenon seen under normal circumstances. To this differentiation I am inclined to attach considerable weight.

Clearly, in respect of the affective seizures, a mechanism of *general motor inhibition* comes into play, in whole or in part; its outward manifestation ranges from mere trembling at the knees to temporary abrogation of the functions of standing, moving, speaking; whether this universal inhibition closures motility from the side of innervation or of tonus, or both, is, as I have pointed out, difficult to determine and in one sense relatively immaterial. My examination of the physiological state during one of these attacks has shown the outfall of tonus to be so absolute as to cause disappearance of the knee-jerks. Where is this

centre of tonus, whose inhibition leads to universal atonia? Is it labyrinthine, cerebellar, mesencephalic, or pre-mesencephalic? The toneless sinking to the ground is distinctly reminiscent of the effect on the boxer of a blow on the point. To quote Sherrington [76]: "The 'knock-out blow,' where the lower jaw conveys concussion to the otocyst, reduces in a moment a vigorous athlete to an unstrung bulk of flesh whose weight alone determines its attitude, if indeed a reactionless mass can be described as possessing attitude at all." Is there loss of deep reflectivity in the minutes that follow the knock-out? Unfortunately I can find no observations to aid in determining the question. Certain it is that consciousness is not by any means always lost, so that the parallel between this state and that of affective cataplexy is sufficiently close physiologically. If merely for the sake of discussion we postulate a labyrinthine origin for the somatic symptoms, and assume the continuance of integrity of the volitional motor system which nevertheless is incapable of overcoming the absolute atonia of the moment, by what mechanism does emotion affect the labyrinth or its central representation? It is conceivable that we have here through disease stumbled upon a biologically old automatism, a defence-reaction in which immobility is of advantage to the frightened animal, "struck all of a heap." If this should seem a feasible speculation, why is the mechanism activated by the act of laughter in so many instances? Hysterical laughter sometimes covers fear, yet the explanation is surely insufficient otherwise.

In the attacks of "sleep," further, it is permissible to see a *generalized inhibition of muscular activity* (on a previous page I have already pointed out that in numerous recorded cases only the appearance of sleep is exhibited, the fundamental feature being inability to move a single muscle). I consider, therefore, that the clue to the phenomena of narcolepsy is to be sought in the direction of generalized muscle- and tone-inhibition. It remains for future study to elucidate the mechanisms by which sensory or affective excitations inhibit instead of arousing motor activities, as well as those through which "sleep" and trance-like states develop "spontaneously."

The problem of pathogenesis, however, may be regarded from another angle. If we take a constellation of nerve-cells at any given neural level, it is theoretically capable of being activated by excitations from the same or from a superior or inferior level, and presumably also of being inhibited. One can consider the sleep mechanism X as being capable of excitation by stimuli of a psychical or psychopathological

kind, A; or by local direct action at its own level owing to the existence of local conditions, B; or by excitations derived from lower levels, C. The formular combinations X × A, X × B, X × C will represent states not differing materially in their outward clinical manifestations, but their respective pathogeneses will be of differing rank. To take an obvious clinical analogy, that of torticollis; this purely symptomatic condition may be pathogenically a tic (psychical), a spasm, an occupation neurosis, a paralytic reaction, a reflex neuralgic reaction. So one narcoleptic state may closely resemble another in externals, but be distinguished pathogenically by the source of its excitant element. In a given instance it may be the outcome of excitations of a psychological order, in others it is a sequel to local conditions of the same level, or to impressions extending from lower levels. And there is no reason to doubt that in the psyche itself there may be levels of function, so that the pathogenic possibilities are correspondingly increased.

Only in some such way as this can we find a possible interpretation for the diverse narcoleptic symptoms of the psychoneurotic, the post-encephalitic, or the sufferer from cerebral tumour, as well as of the patient whose case is classed as spontaneous or idiopathic.

Conclusions.

At various points throughout this discussion, the conclusions to which it appears to lead have been briefly indicated, and further recapitulation is unnecessary. Their tentative nature, however, must be fully admitted.

With unfamiliar conditions like the "narcolepsy" which some persist in calling a "disease," attempts at identification and at nomenclature should come last, not first. The many analogies which various narcoleptic syndromes offer to those usually termed epileptic are here examined without prejudice; differing in outward manifestation, they none the less are such as to suggest that a more or less similar physiology lurks behind each. What precisely this is, however, it is impossible at present to determine.

Again, narcoleptic phenomena on analysis turn out to exhibit close affinities to those not only of epilepsy but also of catalepsy, while it is also found that "sleep" is a term used with vagueness and indeed with inaccuracy in respect of many of the symptoms classed as narcoleptic. The suggestion is that greater significance (from the viewpoint alike of ætiology and of pathogenesis) attaches to the toneless, power-

less, motionless awareness of numerous cases (both in "sleep" attacks and in cataplectic attacks), than to actual sleep.

It is useless to force our cases of "narcolepsy" into the category that seems suited to them unless they fit comfortably; the initial task of minute clinical observation and investigation is still far from reaching its end.

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Proceedings of the Section of Neurology of the Royal Society of Medicine.

President—Edwin Bramwell, M.D.

Clinical Meeting held at the Maida Vale Hospital, on Thursday, November 10.

(1) *Epilepsy with Hemianopia.*

By W. G. Wyllie.

Florence C., aged 45, married. One living child; one died, aged 1 week, from diarrhœa: no miscarriages.

Complaint.—Attacks of drowsiness, with and without loss of consciousness. These commenced at the age of 13. The attacks then lasted about five minutes, during which she could move and hear others speak, but could not speak herself. She had about two attacks a day.

At the age of 17, had "inflammation of the brain" with pyrexia and loss of consciousness. There was no diplopia and she was not sleepy. Afterwards fits ceased until the age of 32.

Since aged 32 she has had attacks resembling *petit mal* in nature, controlled by luminal. There is always an aura of "feeling something in the throat," but no visual disturbances.

Twelve months ago noticed that she did not see the last two words of a line when reading, and that she tended to walk into things on the right side.

Her memory has been deteriorating during the last three years. No other symptoms are present.

Signs.—Very slight rotary nystagmus to both sides, not constant but definite when present. Right homonymous hemianopia present. Slight weakness of right side of face. No other physical signs.

Diagnosis.—Epilepsy and hemianopia.

X-ray of skull (stereoscopic). No bony changes to be made out.

Blood: Wassermann reaction negative; *Cerebro-spinal fluid*: Wassermann reaction negative; protein, ·035 per cent.; cells 2; Lange, 0001000000.

(2) *Epilepsy with Hemianopia.*

By C. Worster-Drought.

F. B., aged 45, complains of seizures and defective vision towards the right.

History.—The first fit occurred four years ago and consisted of sudden loss of consciousness, followed by some degree of aphasia and right hemiparesis,

lasting for one week. The second attack occurred in August, 1926, but was milder in character. Since this date he has had eight further attacks, each one followed by dysarthria and weakness of the right arm and leg of one or two days' duration.

The attacks are preceded by a short aura of giddiness, and during the attack it is stated that he is cyanosed, that the right arm and leg twitch, and that he froths at the mouth; the tongue has also been bitten. No history of venereal disease or head injury, and no previous illnesses of importance.

Physical examination.—Pupils equal and react to light and accommodation; right homonymous hemianopia; fundi normal; other cranial nerves normal. Some loss of joint sense in right fingers and astereognosis right hand; sensation otherwise normal. Arm-jerks moderate and equal; abdominals brisk and equal; knee-jerks equal; right ankle-jerk slightly greater than the left. Plantar reflexes flexor. Occasional involuntary movements of the right hand, especially flexion-extension of right thumb, and some inco-ordination of the right hand; the left hand and both legs are normal. Other systems normal. Arteries not thickened and blood-pressure 150 systolic, 90 diastolic.

The Wassermann reaction is negative in both blood and cerebro-spinal fluid, and the latter normal in all respects. X-ray of skull shows no abnormality.

Dr. HARRIS said that the date of onset of the hemianopia in Case 1 was uncertain. He had seen the association of epilepsy with hemianopia in a case of chronic syphilitic pachymeningitis. He had also seen transient hemianopia lasting up to twenty-four hours, associated with epilepsy in cases of lesions of the cuneus.

Dr. PARKES WEBER hazarded the diagnosis of tuberose sclerosis; although there was no adenomum sebaceum, he pointed out that there were numerous nævi over the face.

Dr. CLOAKE pointed out a definite objective diminution of sensibility over the face and suggested the possibility of pituitary disease in Case 1.

Mr. CAIRNS emphasized the attack of drowsiness in Case 1, and suggested the diagnosis of tumour of the tuber cinereum.

(3) *Case of Peripheral Neuritis, followed by Thrombo-angeitis Obliterans with Gangrene.*

By A. FEILING.

Mrs. T., now aged 48, was first admitted to hospital in April, 1925, for weakness of the legs. She then stated that her feet suddenly "went to sleep" while in bed one night, and that since then (January, 1925) she had had pains in the feet and gradually increasing weakness of the feet and legs. Examination showed a definite weakness of the movements of both feet and ankles, with partial foot-drop and a steppage gait: the ankle-jerks were lost, while the knee-jerks were retained; there was tenderness of the calves and slight loss of sensation for light touch over the distal parts of both feet and legs. No other

abnormality, except that both hands and both feet were always cold. The Wassermann reaction was negative and the cerebro-spinal fluid normal. No cause for the neuritis was found. She gradually improved and was discharged in July, 1925.

She was readmitted in October, 1926, with the symptoms of Raynaud's disease in both the hands and feet. The ankle-jerks were still absent. In July, 1927, she had influenza, followed by grangrene of the left second and third toes, and on July 12 she was again readmitted.

The skin over all the toes was glazed and blue, and both feet were cold; the second and third toes of the left foot showed early gangrene, being purple in colour and stone cold. Pulsation was felt in both dorsalis pedis arteries and in the posterior tibial arteries. The ankle-jerks were still absent on both sides, but otherwise there were no abnormal neurological signs.

On July 18 Leriche's operation was performed on the left femoral artery in Scarpa's triangle. The artery was small in calibre: alcohol was first injected into the sheath, which was then stripped off over a length of $1\frac{1}{2}$ in.

By July 22 definite gangrene had spread to the sole and dorsum of the foot, and on September 1 amputation through the middle of the leg had to be carried out. The stump has healed slowly. About September 18 gangrene began in the toes of the right foot. This has been treated conservatively, and a line of demarcation has gradually formed without further spread of the gangrene.

Dr. F. PARKES WEBER thought that the gangrene in the two lower limbs was due to thrombosis in small arteries complicating angiospastic disturbance of Raynaud's type. He therefore suggested the diagnosis of "peripheral neuritis, followed by Raynaud's angiospastic phenomena and gangrene."

Professor BRAMWELL inquired whether muscular wasting ever followed occlusion of the femoral artery.

Dr. PARKES WEBER replied that such an occlusion might be followed by two types of wasting: (1) due to disease; (2) due to the same cause as the original femoral thrombosis.

Dr. CLOAKE asked for the experience of the Section on the result of Leriche's operation in Raynaud's disease. He had observed one case which benefited considerably.

Dr. CAWADIAS' experience was that the results of Leriche's operation in Raynaud's disease were good in 50 per cent. of the cases. In one case of true thrombo-angeitis obliterans the result was bad, but there was a definite constitutional factor present. Of cases of obliteration secondary to infective arteritis (following influenza or typhoid), he had had no personal experience, but surgeons had told him that local procedures may benefit when the exact level of the occlusion was known. This level might be determined by means of oscillometry.

Dr. HARRIS referred to a case of syphilitic cerebral thrombosis followed by chronic thrombo-angeitis obliterans, in which a bilateral Leriche operation had been of extreme benefit.

(4) *Chorea of Huntington's Type—Possibly Post-encephalitic in Origin.*

By C. WORSTER-DROUGHT.

C. J. C., aged 29, complains of general weakness, slowness of movement, rapid fatigue on exertion and severe headaches.

History.—At the age of 20, while serving in the Expeditionary Force in Egypt during August, 1918, he developed a pyrexial illness accompanied by lethargy. No definite diagnosis appears to have been made; malaria was considered, but a report states definitely, "No malaria parasites found." Several relapses accompanied by pyrexia occurred from this time until his return to England in February, 1920. Following this, he developed the symptoms of which he complains, which have since become progressively worse.

Physical examination.—Pupils equal and react normally: optic discs normal: poor convergence of eyes; facial movements slow, with some right-sided weakness and labial and lingual tremors. Voice somewhat tired and monotonous. Abdominal reflexes normal; knee- and ankle-jerks brisk and equal; plantars flexor. Some degree of general muscular rigidity, incoordination of arms and a tendency to festination.

From time to time he exhibits atheto-choreic movements of the head and limbs. All movements are very slowly and deliberately performed, and frequently preceded by slow, writhing choreiform movements of the arms and trunk or slow gesticulations, particularly on rising from a chair or beginning to walk. Heart, lungs, and other organs normal. Mentally, he exhibits slow cerebration and some dullness, and frequently falls asleep by day when sitting.

Dr. CARMICHAEL suggested the diagnosis of hysteria.

Dr. R. M. STEWART agreed that one had to exclude an element of hysteria from the gait.

Professor BRAMWELL said that the appearance of the tongue suggested post-encephalitic Parkinsonism.

Dr. ADIE said that the whole attitude of the patient, together with his disordered respiration, was suggestive of encephalitis.

Dr. WORSTER DROUGHT said that hysteria had been considered as a diagnosis, but that treatment accordingly had been of no avail.

(5) *Facial Hemihypertrophy: Neuro-fibromatosis; Kyphoscoliosis; Compression Paraplegia.*

By W. RUSSELL BRAIN.

Previous history.—N. S., aged 26, is said to have been normal at birth. Enlargement of the right side of the head and deformity of the spine have gradually developed since the age of 2. The vision of the right eye was lost at the age of 10. Seven weeks ago he first noticed weakness of the lower limbs, and three weeks later he was admitted to the London Hospital with paraplegia and acute retention of urine. The family history is normal.

Present condition.—His intelligence is normal. The whole of the right side of the skull is larger than the left, with special prominence of the temporal crest, the maxilla and the mandible. There is hypertrophy of the skin and subcutaneous tissues of the right side of the face, leading to occlusion of the right palpebral fissure and the right external auditory meatus. The right eye exhibits secondary glaucoma and staphyloma. There is hypertrophy of the gums on the right side, of the right half of the hard palate, and of the right side of the tongue, the papillæ of which are enlarged. The bones of the right upper limb are slightly larger and longer than those of the left.

There is a chain of neuro-fibromata above the right zygoma. The cutaneous pigmentation ranges from slight freckling of the face and neck to coffee-coloured patches 2 or 3 in. in diameter on the left knee.

There is extreme kyphoscoliosis and "pigeon breast." The cranial nerves and upper limbs are normal. There is marked weakness of the lower limbs, the right being more affected than the left, with extensor spasticity and bilateral extensor plantar responses. Relative anæsthesia to cotton-wool over both lower limbs: relative analgesia to prick over both sides of the trunk below the nipples and over the left lower limb; and some loss of postural sensibility in the toes, which is more severe on the right side than on the left. There is now only slight difficulty in beginning micturition. Radiographs revealed thinning of the cranial bones.

Dr. F. PARKES WEBER thought that all the features of the case might be regarded as manifestations of von Recklinghausen's neuro-fibromatosis. The cutaneous pigmentation was typical, but the kyphoscoliosis was of extreme degree. The tongue showed a minor variety of the rare "neuro-fibromatous hemihypertrophy," without a palpable plexiform neuroma. The hanging skin on the right side of the jaw might be called "neuro-fibromatous dermatolysis." There was neuro-fibromatous thickening of the gums on the right side of both the upper and lower jaws, and there was neuro-fibromatous thickening of the periosteum over portions of the skull, with palpable ossification of part of the thickened periosteum. He thought that the diffuse connective-tissue thickening in the skin and periosteum was allied to plexiform neuroma, the connective-tissue increase in the affected parts occurring with only slight neuromatous thickening. He thought that the patient manifested in a lesser degree the changes which were so remarkable in the famous "elephant man" of the London Hospital.

Dr. CRITCHLEY stated that he had seen subperiosteal swellings in at least two other cases of von Recklinghausen's disease, and stated that they had been specifically described as "subperiosteal cysts" by Barney Brooks and Lehmann, in 1924. Pathologically they were due to cystic degeneration occurring in small neuro-fibromata on the subperiosteal nerve filaments.

Dr. ADIE referred to the thickening of the eyelid, which together with an extreme degree of kyphoscoliosis had been the only manifestation of von Recklinghausen's disease in a young patient of his.

(6) *Case of Cortical Epilepsy of Obscure Etiology.*

By E. B. STRAUSS.

W. P., boy, aged 12. Physical development that of a boy of 16. Has suffered from fits on and off since the age of 7 years. Was admitted into the Maida Vale Hospital for the second time last June, suffering from ten to twenty fits daily, which lasted for about a minute. Consciousness appeared to be retained. The eyes were turned to the right, and the head and trunk to the left.

During the fit there was a marked hypotonia of the right arm and leg, and athetoid movements of the right arm and hand. At all times there was a definite right hemiparesis and hypotonia of the arm and leg. The plantar responses were variable. The fits could only be controlled with chloral hydrate gr. xx *ter in die*.

July, 1927, operation to expose left parietal region. Apart from abnormal adherence of the meninges to the cortex and a very dilated vein, nothing abnormal was found. The meninges were partially separated and the wound closed. The patient made a slow recovery and remained in a very peculiar torpid mental state for some weeks. He has remained completely free from fits without drugs since the operation, but for some weeks following the operation the right hemiparesis was more marked than before. On discharge he was mentally normal. There was some protrusion of the dura mater through the wound. He has now completely recovered from his right-sided weakness and hypotonia.

Dr. WORSTER DROUGHT raised the possibility of a subcortical venous angeioma with adhesion to the overlying meninges.

Dr. CARMICHAEL suggested the diagnosis of cerebral circumscribed meningitis.

Dr. FEILING said that the surgeon who performed the operation thought the appearances were unlike those of a venous angeioma, and resembled rather a chronic inflammatory process.

(7) *Subacute Combined Degeneration of the Cord. Recent Blood Crisis with Rapid Recovery following Treatment with a Liver Diet.*

By DOUGLAS MCALPINE.

Male, aged 48, attended the Middlesex Hospital in June, 1923, with symptoms of subacute combined degeneration of the cord.

The chief nervous symptoms were: (1) Slight paresis both lower limbs; (2) absent knee-jerks; ankle-jerks feebly present; (3) vibration sense diminished in lower limbs; (4) plantar responses flexor.

Blood-count.—Red blood-corpuscles, 2,900,000; hæmoglobin, 80 per cent.; colour index, 1·4. No nucleated red cells. Wassermann reaction negative.

January, 1924.—Neurological signs as before. Red blood-corpuscles, 3,900,000; hæmoglobin, 80 per cent.; colour index, 1·0.

July, 1924.—Fractional test meal showed total achlorhydria; knee-jerks and ankle-jerks absent. Treatment with full doses of dilute hydrochloric acid begun.

January, 1925.—General condition had improved; working full hours in a shop. Red blood-corpuscles, 4,300,000; hæmoglobin, 84 per cent.; colour index, 1·0. During 1925 and 1926 condition remained satisfactory.

May, 1927.—Severe blood crisis. Treated at home. Blood transfusion necessary. Patient not seen again until September 23. He stated that progress had been very slow. Blood-count (September 23, 1927): Red blood-corpuscles, 2,150,000; hæmoglobin, 50 per cent.; colour index, 1·2. Nucleated red cells, 1·5 per 100.

September 9, 1927.—Liver treatment begun: ½ lb. per diem.

October 15, 1927.—Patient stated he felt much better since taking liver. Appetite had returned, palpitation had disappeared, and that he felt as well as before his relapse in May. He had returned to work, and could walk a mile or so without fatigue. Blood-count: Red blood-corpuscles, 3,040,000; hæmoglobin, 54 per cent.; colour index, 0·9. No nucleated red cells seen.

November 4, 1927.—Improvement has continued. Right knee-jerk feebly present, left absent. Left ankle-jerk feebly present, right absent. No paresis in legs. Plantars remain flexor. He has gained 9 lb. in weight since liver treatment was begun. Blood-count: Red blood-corpuscles, 3,870,000; hæmoglobin, 82 per cent.; colour index, 1·1; megalocytosis is present.

The knee-jerks are now obtainable; the right plantar is extensor, and the left one flexor. He also quoted two other cases of subacute degeneration treated by liver extract, and emphasized the importance of treating dental sepsis, and also of allowing for spontaneous remissions.

Clinical meeting held at the National Hospital, Queen Square, Bloomsbury, on Thursday, January 12.

(1) *Thyroid Neoplasm with Secondary Deposits in Skull.*

By J. S. COLLIER.

E. E., male, aged 59.

This patient gives a history that fifteen years ago a swelling appeared in his neck. It was painless and grew slowly but steadily. Five years ago the swelling had become so large that he was dyspnœic and had difficulty in swallowing. He was operated on and most of the thyroid gland was removed. He improved and returned to work.

Four years ago he noticed a small lump in the right parietal region. This has grown slowly but steadily and is now tender.

In August, 1927, he again began to have difficulty in swallowing. His breathing was now worse when he was lying flat.

General condition.—Slight stertor as he lies in bed. Gross dyspnœa on exertion.

Cranial nerves.—Pupils unequal, right 3½ mm., left 2 mm. They both react briskly to light and on convergence. Visual acuity $\frac{6}{12}$ both eyes. Fundi normal. The right palpebral aperture a little wider than left. Right eye displaced slightly down and up.

Motor and sensory systems normal.

Reflexes.—Normal, except that the left ankle-jerk is absent, but he had sciatica sixteen years ago.

Skull.—There is a rounded hard mass in the right parietal region which rises smoothly from the general skull surface. There is a soft area at the top of it which is tender to pressure.

Cardiac system.—The apex beat is 5½ in. from mid-line. Diastolic shock over the aortic area. There is some extension outwards of dullness in the second right space. Heart sounds: there is a short harsh systolic murmur accompanying the first sound over the aortic area. The aortic second sound is ringing and accentuated.

Blood-pressure: Left arm 146/78; right arm 158/92.

Cerebro-spinal fluid: Normal.

Dr. WILKINSON said that the first suggestion was that the cranial tumour was gummatous: the spinal fluid and blood Wasserman reactions were both negative, however. It was now suggested that the skull lump was independent of the thyroid tumour.

Dr. FEILING asked what evidence there was that he had ever had an actual neoplasm of the thyroid.

Dr. WILKINSON said that there was no proof beyond the patient's statement to that effect.

Dr. ADIE said he first saw the patient six weeks ago, and he had thought the swelling was a gumma.

(2) *Epilepsy following Contusio Cerebri.*

By J. S. COLLIER.

F. K., male, aged 12.

This boy gives a history that in August, 1925, he fell 17 ft. and struck his left occiput and left mastoid region. He was picked up unconscious. He remained so for ten days. On recovering consciousness he was completely blind for two days—his sight then returned.

There was no bleeding from mouth, nostrils or ears. He remained in hospital three months.

In May, 1926, nine months after the accident, he had a fit. He fell suddenly in the street and his limbs were convulsed. His lips went black and he frothed at the mouth. He wetted himself but did not bite his tongue. He remained unconscious for ten minutes.

In November, 1926, he had another fit of the same type. In March, 1927, he had another attack—this attack was preceded by an elaborate visual aura. He states that in the left visual field he sees people walking past him. They move from left to right across the field and he can still see them if he closes

the right eye. He can distinguish men from women in the crowd, but cannot tell the colour of their clothes. Their faces are fairly clear, but he does not think he would recognize them in real life. He has never heard them speak—they pass for from three to five minutes. Then his hands and fore-arms begin to jerk and he falls unconscious to the right.

On October 14, 1927, he had another attack at school. This was preceded by a similar aura and the attack then followed its usual course. Every attack has been succeeded by vomiting and headache.

He wetted himself in the first one only, and has not bitten his tongue in any of them. Between the attacks he has been in normal health.

Cranial nerves.—Pupils unequal, right 4 mm., left 5½ mm. The left reacts briskly to light, and on convergence. Right: There is the tiniest flicker to bright direct and consensual light. The right pupil reacts well on convergence. Fundi normal. Left visual acuity $\frac{6}{6}$, right $\frac{6}{9}$. External ocular movements normal. No nystagmus, diplopia, or en- or ex-ophthalmos. Right half of face acts a little better than left in both upper and lower halves.

The motor and sensory functions and all the reflexes are normal.

Respiratory system normal.

X-rays of his skull suggest a fracture in the right occipital region.

Sir J. PURVIS STEWART said an "occipito-temporal fracture" would be a correct designation: the skiagram showed that the fracture involved the temporal bone, and the character of the visual aura suggested a temporal site. Absence of a quadrantic field defect suggested a cortical lesion.

Dr. C. P. SYMONDS wondered whether the site of the fracture was independent of the site of the cerebral contusion. He had thought that the lesion was in the neighbourhood of the thalamus, and that the hallucinations were due to disturbance in the optic radiations rather than in the temporal lobe.

(3) *Polycythæmia with Cerebral Thrombosis.*

By C. M. HINDS HOWELL.

S. N., aged 29, suddenly developed a right-sided paralysis three months ago. It came on while he was at work, and was unattended by loss of consciousness. This attack lasted only a few minutes, and he felt quite well afterwards. A second attack followed fifteen minutes later, and in this one his speech became affected. Four more transient attacks occurred before the evening. The following day he had another severe attack, which was followed by a permanent right-sided weakness.

The right hemiparesis is now improving; speech is almost normal. Spleen enlarged. Blood-pressure 135. Red blood-count 12,000,000.

Dr. CRITCHLEY said he had had no experience of a cerebro-vascular disorder associated with polycythæmia.

Dr. SYMONDS states he had seen one case of a vascular disorder in this disease. Later the patient had died from some intercurrent disease, and an old hæmorrhage into the brain was found at the autopsy.

Dr. F. PARKES WEBER regarded the case as a typical one of erythræmia (splenomegalic polycythæmia). Cerebral thrombosis had been early recognized as an occasional complication (e.g., case of Hutchinson and Miller, 1906). He thought that the white blood-cells should be counted and a differential count made in every case of erythræmia, so as to distinguish the cases bordering on myeloid leukæmia—the so-called cases of "erythro-leukæmia" (Guglielmo) and "subleukæmia erythræmia" (Aubertin). (Compare F. P. Weber and O. Bode, "Erythro-Leukæmia and Blumenthal's Disease," *Med. Press*, London, 1927, vol. clxxv, p. 174.)

If a trial of phenylhydrazine treatment were made, care should be taken to discontinue the drug when the erythrocyte count fell as low as 5,500,000, as a further fall was likely to occur during seven to ten days following the termination of the treatment.

(4) *Schilder's Disease* (?).

By E. A. B. PRITCHARD.

A. A., male, aged 12. This boy, who is the youngest member of a family which shows a tendency to mental deficiency in the males and average intelligence in the females, developed normally until he had an accident at the age of 3½ years. Since this time there had been a gradual deterioration in several respects. His speech became slurred, and is now limited to meaningless sounds; his hearing has deteriorated, so that it is now doubtful whether he can hear at all, and he certainly cannot understand spoken words, while his general movements have become awkward and irregular, with increasing spasm and athetoid movements of the trunk and limbs. One aunt had become prematurely deaf; his mother was very deaf. The patient was the sixth of seven children; his three sisters were of very bright mentality; the three brothers were extremely dull. The diagnosis of an unusual variety of Schilder's disease had been suggested.

Dr. SYMONDS referred to a case of familial Schilder's disease he had shown one year ago. Ferraro had since then published three cases of familial Schilder's disease. Krabbe's description of a variety of diffuse cerebral sclerosis presented the pathological picture of this same disease. This particular patient corresponds with the description of the Pelizaeus-Merzbacher syndrome, a disease in which males only were affected. The query again arises as to whether Pelizaeus-Merzbacher's disease and Schilder's encephalitis were not one and the same disorder.

(5) *Palato-Laryngeal Nystagmus.*

By S. A. K. WILSON.

A. B., aged 53, was admitted for weakness of his left arm and leg, failing sight and bad memory, of twenty-four months' duration. Sudden onset twenty-four months ago, on waking one morning, of weakness of his left arm and leg, from which he made a gradual recovery. Three months ago he suffered with

severe headache, and a day or so later became suddenly blind whilst in the street; this persisted for two months and sight has since gradually returned.

A week later he had a "stroke," heralded by a fit in which there were right-sided convulsions. Speech was lost for three days, but there is no history of subsequent motor weakness. Memory has recently become very bad.

Physical examination.—A patient of poor mentality; memory bad, co-operates poorly but is well-orientated. Speech is indistinct. His pupils are oval, and left greater than right, but both react normally. The retinal vessels are tortuous, with indentations of veins by arteries and pigmentary changes at the maculæ. Palatal nystagmus is present. This is regular in rate; on admission the movement was at the rate of about 144 per minute, but is now slower, about 116 per minute, and takes place in a vertical direction. In addition there is a horizontal nystagmoid movement of the left superior constrictor muscle, and of the left posterior faucial pillar. There is slight horizontal nystagmus of the right superior constrictor muscle. The palate moves poorly, but equally and symmetrically: the palatal reflex is normal. The tongue is protruded to the right and is tremulous but not wasted. The left limbs are weak and the tone of their muscles is increased, but there are no sensory changes. The reflexes are exaggerated but equal in both upper limbs. The abdominal reflexes sluggish in the upper and absent in the lower quadrants. The right and left knee- and ankle-jerks are exaggerated and there is ankle-clonus on both sides, but the plantar responses are flexor. Some precipitancy of micturition exists. The heart is not enlarged, but the aortic second sound is accentuated. Blood-pressure is 190 112.

Examination of vocal cords shows well-marked twitchings of the left arytænoid and left vocal chord, synchronous with those of palate. There are no involuntary movements on the right side.

Examination of the cerebro-spinal fluid reveals very slight increase in protein content only. Wassermann reaction is negative. The Lange test shows no change.

Sir J. PURVES STEWART suggested that the term "myoclonus" was preferable to "nystagmus" to describe the involuntary movements present in this case.

Dr. CRITCHLEY stated that this variety of movement appearing in the palate, pharynx and larynx was not very uncommon in cerebro-vascular disease. In view of the diffuseness of the lesion, it was difficult in such to correlate the pathological and clinical observations. Palatal "nystagmus" had, however, been seen in numerous cases of focal lesions, such as in tumour of the cerebellar vermis (as in the first case recorded), in focal softenings in the dentate nuclei or in the pons. Dr. Wilson had spoken, in 1921, of a case of palatal nystagmus associated with tumour of the corpora quadrigemina. Dr. McAlpine and Professor Lhermitte had described choreic movements of the face, palate, and larynx which they associated with neo-striatal disorder. In the most recent work on this subject, Signorelli had implicated the substantia reticularis.

(6) *A Case showing Tonic Innervation.*

By C. P. Symonds.

L. H., aged 15, first came under observation at the age of 12, in November, 1924. The mother stated that, having been previously healthy, a year before it was noticed that she was using her left hand more than her right, and that she tended to swing her right leg as she walked, wearing out the toe of her right boot. For the past month she had been complaining of headaches when waking in the morning.

On examination, there was slight weakness of the right side of the face, and the right arm and leg, with diminished abdominal reflexes on the right side. The plantar responses were both flexor. Nystagmus was noted on conjugate deviation to the left. The Wassermann reaction in the blood was negative. X-rays of the skull were negative. X-rays of the chest showed a large gland at the right root and some rounded opacities arranged in streaks, suggesting some infiltration of the lungs.

She was admitted to hospital, where no further physical signs were discovered. Her visual fields were full on the perimeter. There was no aphasia.

On the suspicion of a tuberculoma in the brain-stem she was discharged to a convalescent home at Broadstairs. There she remained for six months.

On her return in June, 1925, she attended for re-examination. The weakness and physical signs remained as before, but she was free from headaches, and she returned to school.

Two years later, in June, 1927, she began to complain of frontal headaches. After this had continued for a fortnight, she was seized one morning while at stool with sudden severe pain in the head, vomited, and within a quarter of an hour became unconscious.

Five and a half hours later she was admitted to Guy's Hospital in a semi-comatose condition. There was well-marked cervical rigidity and bilateral Kernig's sign. The abdominal reflexes were with difficulty obtained on the right side. The right plantar response was extensor. The right big toe occasionally went into a position of extreme dorsiflexion with the foot in a position of plantar flexion, this posture being strongly maintained for half a minute at a time.

Lumbar puncture gave an evenly blood-stained fluid with the characteristic appearances of a spontaneous subarachnoid hæmorrhage. This was not recognized as such at the time, and further lumbar punctures were done on the subsequent day and again two days later under the impression that the hæmorrhage was accidental. On each occasion the appearances were the same, except that with the passage of time the supernatant cerebro-spinal fluid standing in the test tube became more definitely golden-yellow in colour.

Four days after admission she was able to co-operate in a complete examination. There appeared to be some slight difficulty in speech production. There was weakness of right face, arm and leg, but no sensory disturbances.

It was noted that "when the fingers of the right hand are clenched the patient has great difficulty in initiating relaxation, and attempts to start this movement with the aid of her left hand."

Seen on the fifth day of her illness, she showed bilateral papillœdema of moderate degree, with a few small retinal hæmorrhages, in addition to the other physical signs already described. The papillœdema continued to increase during the next three or four days. Her temperature was raised for the greater part of a week. The right-sided weakness improved, and the right plantar response became flexor.

On July 8 Dr. Ironside noted Meyer's reflex present on both sides, exaggerated on right. "On stroking the palm in the region of the carpo-metacarpal joints on the right, the fingers go through two or three movements of alternate flexion and extension, and the hand moves slightly towards the stimulus applied to the palm."

The difficulty in relaxing the right-hand grip improved. The patient's general condition rapidly became normal, and she was discharged on August 8.

Since then she has been completely free from headaches, and in good health, apart from the continued slight hemiparesis and "tonic innervation" of right hand and foot.

Present condition.—General health is good and physical development and mental state normal for her age. She is right-handed: no left-handedness in family. No speech defect is discovered on receptive or productive sides. The visual fields are normal to rough tests. Optic discs are both pale with clear-cut eyes. (Probably consecutive atrophy following the papillœdema noted above.) The pupils are equal and central, the right a little irregular: they react normally to light (direct and concentral) and on accommodation. The ocular movements are full with nystagmus, fine to right, coarser to left. The corneal reflexes are present and equal. There is weakness of the right lower half of the face, equally present in voluntary and emotional movement. The palate moves in mid-line and the tongue is protruded centrally. In the right upper limb individual movements of the hand and fingers are good, but power is less than on the left. Consecutive quick movements, e.g., piano-playing, are slow. There is some spasticity of the flexors of wrist and fingers. In all positions of the trunk and limbs the right hand is held slightly flexed and adducted to the ulnar side. This is perhaps most apparent when the hands are held above the head. When the hands are held outstretched with fingers spread, the same postural abnormality is to be observed, associated with irregular jerky movements of adduction and abduction in the fingers.

When she grasps the observer's hand firmly there is increased flexion and ulnar adduction at the wrist, with some difficulty in relaxing the grasp owing to persistent tonic contraction of the flexors, which can be seen and felt in the fore-arm. The same difficulty is much more marked when she makes a fist. It may be as much as half a second before she can relax the flexors, in spite of powerful contraction of the extensors during this time. The effort to open the fingers is associated with increased flexion and adduction of the wrist. After

she has opened and shut her hand two or three times the difficulty in relaxing the flexors becomes less, but is usually as well marked as ever after a rest. Other movements of the right upper limb are normal, with no difficulty in relaxation. The triceps, biceps and supinator reflexes are all more easily obtained on the right than the left. Stroking the palmar surfaces of the right fingers over the proximal phalanges, from ulnar to radial border, occasionally provokes a slight adduction of the thumb, not to be obtained from the other hand. No postural reactions of the type described by Riddoch and Buzzard could be evoked from the right upper limb, nor could any alteration of postural tone be demonstrated by alteration of the position of the head.

In the trunk no weakness can be observed. The abdominal reflexes are all present and equal.

In the lower limbs there is slight spasticity and weakness of the flexors of the right ankle and toes. The patient states that she has observed the same kind of difficulty in relaxing the toes as the fingers. This can be demonstrated clearly by asking the patient to grip the observer's finger tip under her toes. There is a persistent contraction, apparently of the long flexors of the toes, which involves flexion of the ankle as well. The duration of the tonic after-contraction here is much shorter than in the case of the fingers. There is the same tendency eventually to obtain release by momentarily increasing the movement of flexion. The action is similar to that instinctively used to get rid of a cramp, though in the patient's case there is no pain. Other movements of the lower limbs are normal. The knee- and ankle-jerks are present and equal on the two sides; the plantar responses are flexor.

The sphincters and gait are normal.

Blood-pressure is 95 and 55.

No abnormalities have been detected in head or spine, or in any of the other systems.

Dr. CRITCHLEY stated that the description of the phenomenon, as given on July 8, suggested rather a "grasp reflex." The two phenomena of forced grasping and tonic innervation both occurred as constituents of the syndrome of the anterior cerebral artery. He suggested, therefore, that the lesion in this case resulted from an aneurysm of the anterior cerebral artery.

Dr. SYMONDS considered this the most reasonable explanation.

(7) *A Case of Familial Nervous Disease (Disseminated Sclerosis).*

By C. P. SYMONDS.

A. W., aged 24, gives a history of difficulty in walking since 1921, attributed to a fall in January of that year, after which she felt numb all down the left side.

She first came under observation as an out-patient at Guy's Hospital in February, 1922, when she showed left hemiparesis with indefinite sensory disturbance and nystagmus.

Later in the same year she began to have attacks, lasting two or three

minutes, in which she becomes completely disoriented in space, and has a feeling of impending disaster. She has never lost consciousness in these attacks, but cries out in fear. They come on suddenly at any time and without warning. She has never had one when alone.

In July, 1925, she first noticed diplopia, especially on looking to the left. At present she shows the following physical signs :—

Mentally she is somewhat childish. Speech is not abnormal.

There is a high degree of myopia, but the optic discs are normal.

The right pupil is larger than the left, but the pupillary reactions are normal. There is fine nystagmus on conjugate deviation to right and left. Diplopia is sometimes present when she looks to the left, but there is no demonstrable weakness of ocular movements. The left lower face is slightly weaker than the right. The remaining cranial nerves are normal.

There is no wasting or weakness of the upper limbs, but a tremor of the intentional type is present on both sides, being more marked on the left. Of the lower limbs the right is more powerful than the left. There is gross inco-ordination in the performance of voluntary movements, such as the heel-knee test.

There is no demonstrable organic loss of sensation, but there is a functional hemihypalgesia.

Triceps, biceps and supinator jerks are present and equal on the two sides. Of the abdominal reflexes the right lower only is obtained. Knee- and ankle-jerks present and equal. No ankle- or patellar-clonus. Plantar responses are both extensor.

There are no physical signs in the other systems.

The Wassermann test is negative in blood and fluid. The cerebro-spinal fluid is clear and colourless : cells 4 per c.mm., 5 per cent. large mononuclears ; total protein 0·1 per cent. ; Nonne-Apelt ++ ; Pandy + ; Lange 0001101000.

Family history.—A brother died in 1926, at the age of 32, of a disease diagnosed as disseminated sclerosis. The symptoms began in 1921 with diplopia. In 1922 he had weakness of the legs and tremor of the hands, and it is recorded that he then had nystagmus, weakness of the right hand grip and an extensor plantar response on the right side. A note in 1923 records nystagmus and strabismus, spastic-ataxic gait, scanning speech, gross inco-ordination and intentional tremor.

One sister died, aged 35, two years ago. The date of onset of her disease is unknown, but in January, 1925, she had diplopia, exaggerated reflexes, double extensor responses and absent abdominal reflexes. The Wassermann reaction was negative in the spinal fluid and the gold curve normal.

This case had been reported in the *Lancet*, 1927, ii, 701, under the heading of Familial Disseminated Sclerosis. The suggestion is made that, without pathological evidence, this diagnosis must be regarded as doubtful, and that the case belongs to the group of familial nervous diseases of which the familial cerebellar ataxies and familial spastic paraplegia may be taken as examples.

Dr. WORSTER-DROUGHT referred to two sisters under his care with

symptoms resembling disseminated sclerosis. A third sister had died, with symptoms very like disseminated sclerosis.

Dr. ADIE stated that he and Dr. Critchley had recently collected a number of cases of familial disseminated sclerosis. There was at least one case in the literature in which the diagnosis of disseminated sclerosis had been confirmed by autopsy. This case was described by the younger Hoffmann.

Dr. CLOAKE spoke of a similar family under his care. One member had symptoms resembling disseminated sclerosis with the characteristic disc changes. A sister was an even more advanced case; the previous generation also showed a nervous disorder resembling disseminated sclerosis.

(8) *A Case of Dermato-Myositis.*

By J. P. MARTIN.

Patient, Mrs. B., aged 50.

History.—About thirteen months ago began to have difficulty in getting up from a chair and in raising her arms; these disabilities gradually increased; after a few months she was unable to stand, and when sitting had difficulty in holding up her head; for the last four or five months she had been confined to bed and has become thinner.

Present condition.—The skin on the lateral parts of the face, on the chest and shoulders, the right side of the abdomen and the anterior surfaces of the legs, and to a less extent on many other parts of the body, is thickened and bound down to the underlying tissues. The muscles of the shoulders and chest, the biceps brachii, the hamstrings and other muscles are hardened, and in the pectorals, biceps brachii and hamstrings there are contractures; sensation is everywhere normal, and there are no signs of disease of the central nervous system; the cerebro-spinal fluid is normal.

The patient is being treated with fibrolysin, massage and warm baths, and improvement is noticeable in the skin and muscles of the neck.

Gowers first described this condition, and later Oppenheim. The patient had recently shown some improvement.

Professor BRAMWELL referred to a similar case associated with facial hemiatrophy. This combination had also been referred to by Gowers.

Dr. F. PARKES WEBER said that the patient was very decided that the first symptoms were simple weakness, and not stiffness or pain, in the muscles; that was a strong point against the diagnosis of any kind of dermato-myositis or of sclerodermia. He suggested that the case was a rare form of chronic generalized symmetrical multiple peripheral neuritis in which the muscle fibres themselves, as well as the nerves, were involved. The subsequent hardening of the muscles and slight adhesion of the skin might be secondary. There was no obvious alteration in the texture of the skin itself.

The case described by Gowers was that of a woman, aged 36, whose first symptoms were a constant tired feeling and aching pain in the back and around the loins. Both nerves and muscles were thought to be involved—a poly-

neuritis accompanied the polymyositis, but the muscular affection was far more extensive than in case of pure polyneuritis (see Gowers, *Brit. Med Journ.*, 1899, p. 65). The disease might be termed a *chronic symmetrical generalized neuro-myositis.*

Clinical Meeting of the Section of Neurology, March 8, 1928.

Discussion on the Narcolepsies.

Dr. S. A. KINNIER WILSON introduced the discussion by presenting to the meeting an abstract of his paper published on p. 63.

Dr. WORSTER-DROUGHT referred to an article published in the *British Journal of Psychology*, in which he described some of the earlier cases he had observed. At present he has four cases under observation, three men and a woman. In one of these, in which the toneless attacks commenced before the attacks of sleep appeared, he had been able to observe the patient during an attack: his head suddenly dropped forward, his lower jaw fell, and he suddenly became flaccid and powerless. While in this state his arm-jerks and knee-jerks could not be obtained.

He agreed with Dr. Wilson that narcolepsy should be regarded as a symptom-complex rather than as a disease, and it seemed to him that it is closely allied to the condition known as catalepsy.

Sir J. PURVES-STEWART referred to his observation several years ago of attacks of hypersomnia associated with lesions in the neighbourhood of the floor of the third ventricle. He has lately observed excessive sleep following inflation of the third ventricle with air. It seemed to him difficult to explain such paroxysmal phenomena as distinguish narcolepsy by the existence of a stationary lesion or functional disturbance in the brain.

Dr. GRAINGER STEWART said that during the last three years he had observed three typical cases of narcolepsy. One of these was particularly interesting as the narcolepsy developed in a child who was subject to epilepsy. At first while epileptic seizures still occurred, the child began to have typical cataplectic attacks, which were generally brought on by the first taste of food, which he assumed acted as an emotional stimulus. Later both the epileptic and cataplectic attacks ceased and were replaced by periodic attacks of sleep. In one cataplectic attack the knee-jerks were absent, but he could not elicit extensor responses, the plantar reflexes being either absent or indefinite.

Dr. PHILIP CLOAKE offered as an interesting contribution to the subject the record of attacks of sleep and cataplexy described by a girl under his care.

"I am 22 years of age: I first noticed a tendency to sleep unnaturally between the age of 19 and 20; even as I walked along I would fall asleep; several times I did not know even how I managed to get from one end of the work-room to the other; I would suddenly wake with a start and find that I must have been asleep. For at least six months every time I fell asleep while I happened to be in a sitting position, my head would fall back as far as

possible, and then flop right forward for at least a dozen times, and I was quite unable to stop it. As time went on I found that as soon as I lay down in bed I could not move or speak, and if I tried to speak I only made a muffled sound; but if anyone happened to come in the room and spoke I could hear what they said quite well, and if they passed in front of my eyes I could see them though my eyelids felt almost shut. I would have as many as six or seven of these attacks straight off, but each one would last only a minute or so.

"During the last eighteen months these turns have become stronger and it requires a greater effort to pull myself round; I have to make two or three attempts before I can do so, and then when I get up I usually jerk forward once or twice before I can steady myself, but except for feeling a little out of breath I am quite all right.

"I fall asleep at all times, even standing up at work, and I am so bad that I have to go out and talk to someone, or go out in the air before it passes off; and then more often than not during the next half hour I am just as bad. This will keep on perhaps all day, and then next day I only feel it once or twice. After this sleepy feeling passes off my eyes feel very sore.

"When I laugh my legs give way and I have to hold on to anything or anyone near or I should fall; also my eyes seem to roll about and for a minute or so all seems a blank, then everything seems muddled up and out of place, and I have to shut my eyes before I can see things plainly, and generally instead of seeing one I see two; this also happens if anyone takes me by surprise and I turn round quickly. I have to wait a minute or two before I can speak."

One special feature of this case is the curious state of mental confusion that followed the attacks. He was not aware that cataplectic attacks occur in symptomatic narcolepsy, as in that which occasionally follows encephalitis lethargica.

Dr. HENRY COHEN said he had observed eight cases of narcolepsy, in three of which the condition followed attacks of epidemic encephalitis. He was impressed by Dr. Wilson's emphasis on the affinity of narcolepsy to epilepsy, and his own observations tended to confirm this view. In one of his cases from twenty to thirty cataplectic attacks occurred each day, in fact any shock or unexpected noise might precipitate an attack. He had seen a case of congenital spastic diplegia in which a noise might bring on a similar cataplectic attack which was often followed by myoclonic twitchings. This case in many ways resembled that of a woman with congenital syphilis under his care, in whom sudden noises excited epileptic seizures. He had observed true cataplectic attacks, occurring usually after laughter, in a patient who had had epidemic encephalitis.

Dr. GORDON HOLMES referred to two cases of true narcolepsy, one a young man and the other a young woman, which he had seen. Neither of these presented any special features except that the man, whose case has been referred to by Dr. Wilson in his paper, had been subject to epistaxis before the narcoleptic symptoms appeared.

He had the opportunity of making an interesting, and, it seemed to him, an important observation in a middle-aged man, who probably has a slowly growing tumour in the hypothalamic region. He has lately increased greatly in weight and has become subject to attacks of indomitable sleep, lasting ten to forty minutes, from which he wakes up feeling quite natural. In addition he has had a few sudden and unexplained "collapses," or falls on emotion. His symptoms improved very much on the administration of large doses of thyroid and recurred when the dose was reduced. He referred to the case he had described of "Falling on Noise and Surprise" (*Brain*, 1927, **50**, 720), which seemed in certain ways allied to narcolepsy. This boy was from 5 to 14 years of age liable to fall flaccidly on noise and surprise; at the age of 14 years he had an epileptic attack, and since the case was recorded six months ago he has developed frequent and severe epilepsy.

Dr. R. D. GILLESPIE mentioned that in the case which he had observed slight emotional stimuli might excite an attack, while stronger emotions had often no effect. Hot baths also tended to cause attacks of sleep. In some of the attacks of sleep automatic activities might continue for a time, as writing. In one attack he found the knee-jerks absent and could not obtain the plantar reflexes. This case improved for a time under psychotherapeutical treatment, and later on large doses of hyoscine hydrobromate.

Notices of Recent Publications.

Conditioned Reflexes. An Investigation of the Physiological Activity of the Cerebral Cortex. By I. P. Pavlov. Translated and edited by G. V. Anrep. Pp. 430. Oxford: University Press. 1927. Price 28s.

It is several years now since such a noteworthy contribution to physiology, and especially to neurological physiology, appeared in the English language as Professor Pavlov's book. We have been to some extent familiar with the trend and the general results of the investigations to which he and his school have devoted the last twenty-five years, but as almost all the publications on the subject have appeared in Russian and have been widely scattered in various journals, few English readers have had the opportunity of making a first-hand acquaintance with them. We are therefore under a debt of gratitude to Dr. Anrep, who has translated and edited the volume in which Professor Pavlov has collected for the first time the general results of these investigations. The translation itself is distinguished by a clear and concise style which frequently suggests to the reader an original work by a master of our language.

The book takes the form of a series of lectures in which the development of the subject is traced to a succession of general conclusions. In the first chapters we learn of the mode of production and the factors that influence the appearance of conditioned or acquired reflexes; then the various forms of inhibition and the part it plays in the normal activity of the cerebral cortex are illustrated and discussed. The next chapters are devoted to the analysing and synthesising activity of the cerebral hemispheres and to the irradiation and concentration of nervous processes in the cortex. This is followed by a discussion of the essential nature of cortical activity, and of the physiological properties of different parts of the cortex.

From this stage we are presented with a series of more specific problems. In the first place it is shown that, as far as their mechanisms go, sleep and internal inhibitions are the same process, and that when the inhibition does not spread to subcortical centres phenomena identical with those of hypnosis appear. We are here furnished for the first time with a physiological explanation of sleep, and an explanation which will fail to convince few who study these chapters carefully. The psychological hypotheses of sleep and hypnosis that have for long held the field can now be discarded. Other phenomena also which have been regarded as "psychological" receive physiological explanations based on controllable experiments; for instance "suggestion" in man is shown to be one of the simplest forms of conditioned reflex. His rigidly

scientific attitude forces the author to deny to psychology the right to be regarded as a science at all, in fact he insists there is no real line of demarcation between "nervous" and "psychic," the distinction is only arbitrary.

The last seven chapters have a closer relation to clinical neurology. Early in his study of conditioned reflexes Professor Pavlov observed functional disturbances of the nervous system in his experimental dogs which resemble and are obviously closely allied to different types of neuroses met with in man; further investigations have shown that they are due to a conflict between the processes of excitation and inhibition, or to the action of extremely powerful stimuli, and have revealed the means by which they can be successfully treated. The effects of surgical lesions of the cortex on its normal activity as studied by the method of conditioned reflexes lead to an important discussion on the question of localization of function in the cortex, and the form that this localization takes: one conclusion is that his experiments emphatically refute the doctrine of special "association" centres, or of any special area on which the higher functions of the nervous system depend. The final chapter deals with the application of the experimental results to man.

It is impossible to review adequately in a limited space a volume which is not only stored with observations of an entirely new character, but which presents points of view with which few have been hitherto familiar. A mere list of its contents will, however, serve to show that its study is essential to all interested in any branch of neurology.

Travaux et Mémoires. Par Pierre Marie. Tome 2. Pp. 393. Paris: Masson et Cie. Price 30 francs.

The first volume of the reprints of his scientific articles which Professor Pierre Marie is issuing has been already noticed in this Journal. The second number contains a large number of short articles, some of which were published many years ago, others more recently, as well as a few of his longer and more important papers. Among these may be mentioned the original description by Charcot and Marie of "Peroneal Muscular Atrophy," which here appears under the title "Amyotrophie Charcot Marie"; his paper on "Hereditary Cerebellar Ataxy"; that on "Les Lacunaires," or pathological states distinguished by multiple small cavities in the brain; the pathology of Amyotrophic Lateral Sclerosis, and a discussion on the question of localization in the Internal Capsule.

This volume contains many articles which are still important from the pathological, as well as from the historical, point of view, and very few that offer no interest. An index would have made the two volumes more useful.

Beiträge zum Tieproblem. Von J. WILDER and J. SILBERMAN. S. 100. Berlin: S. Karger. 1927. Price M. 8.40.

A critical review of our present knowledge of the nature and pathogenesis of tics is certainly required. This monograph is a contribution to the subject,

but it can scarcely be claimed that it suffices our needs, for on the one hand it includes too many different conditions, as involuntary motor phenomena of organic origin as well as numerous functional or psychogenic disorders; while on the other its authors fail to analyse sufficiently the exact nature and mode of origin of some of the conditions with which they deal. There is, however, an adequate review of the literature on ties, and many interesting points are brought out, especially in the discussion on so-called functional ties, in the study of which they employed a modified form of psycho-analysis. In places there is a tendency to too much theorizing, as in the assumption that mental and emotional activities of relatively high levels are associated with the activity of the corpus striatum, a hypothesis they make to explain the frequent psychial disturbances that accompany ties of striatal origin. The second part of the monograph consists of the case histories and analyses of a large number of patients observed by the authors.

Nerve Tracts of the Brain and Cord. By W. KEILLER. Pp. 456. New York: The Macmillan Company. 1927.

New textbooks on the anatomy of the nervous system appear year by year; some are written from the purely academic standpoint, while others are little more than introductions to elementary neurology. The author of this book professes his aim to be to enable students to study nervous diseases in terms of anatomy, physiology and pathology, and he has certainly produced a work which, if too detailed and voluminous for the ordinary student, will be of considerable use to the post-graduate worker.

In the first section of the book the central nervous system is studied in a series of sections, which are well represented by drawings, first as they are seen by the naked eye and then under the microscope. Owing to this arrangement there is a certain amount of repetition, which, though possibly advantageous in the classroom, adds unnecessarily to the size of the volume. The frequent references to the functions of the structures described will stimulate the student's interest.

In the second section, the motor, sensory, cerebellar and other systems are treated separately, and the functional significance of the different parts of each are discussed. A large part of this section is consequently physiological, and in it the author is on less sure ground and not so up to date as in the anatomical descriptions. The third section on, "Applied Neurology," attempts to explain clinical symptoms in terms of the functions of the parts involved by the lesions that produce them.

Almost all the illustrations are collected at the end of the book; many of them are not original, but the author has shown considerable discrimination in their choice. Unfortunately some of these, as well as his own drawings and diagrams, are reproduced on such a small scale as to diminish their value seriously.

Nasal Neurology, Headaches and Eye Disorders. By G. SLUDER. Pp. 428 with 167 illustrations. London: H. Kimpton. 1927. Price 50s.

The greater part of this book has no interest for the neurologist, while in the sections that deal with symptoms of nervous origin he will find many statements with which he cannot agree, and hypotheses that will astonish him. Several syndromes are described: (1) Vacuum frontal headache associated with eye-strain, the latter being explained by the attachment of the tendon of the superior oblique muscle to the wall of the frontal sinus. (2) Anterior ethmoidal or naso-ciliary neuralgia, characterized by pain in the forehead, between the eyes and along the nasal bones: it is due to a neuritis of the anterior ethmoidal nerve which lies superficial as it enters the nose and is therefore easily involved. (3) Nasal ganglion neuroses, due to the spread of inflammation or of toxins to Meckel's ganglion, which lies in closer relation to the nose than is generally believed. This may take the form of "lower half headache," or pain spreading from the nose and orbits along the base of the skull to the mastoids, occipital region and shoulders; or the "sympathetic syndrome" consisting of vasomotor and secretory phenomena, "very wide in distribution and wondrously complex," and asthma. (4) Sphenoiditis associated with inflammatory or toxic affections of the optic, ocular and trigeminal nerves.

Birth Injuries of the Central Nervous System. Part I: Cerebral Birth Injuries. By FRANK R. FORD. Part II: Cord Birth Injuries. By BRONSON CROTHERS and MARIAN C. PUTNAM. Medicine Monographs, Vol. XI. London: Baillière Tyndall and Cox. 1927.

In Part I of this most interesting work the author attempts a survey of the vexed question of the relationship of birth injuries to the subsequent development of nervous disease. His personal material has been gleaned from an analysis of 200 cases of bilateral spastic paralysis. The literature of the subject has been well investigated and is given in a useful bibliography. The author's conclusions are avowedly imperfect and partake more of the nature of a series of impressions. There is, however, very convincing evidence that the congenital diplegias, which constitute by far the largest group of infantile spastic palsies, are not to be attributable to meningeal hæmorrhage at birth, but are the result of various pathological processes of intra-uterine origin. No final statement could be made as to the relation of intracranial birth injury and chronic hydrocephalus.

The common diffuse meningeal hæmorrhage which is not large enough to cause death apparently leaves no residium in the overwhelming majority of cases. The true traumatic cerebral birth palsies are represented clinically by the hemiplegias, the monoplegias, and the asymmetrical and unequal bilateral spastic paralyses, and have for their pathological basis intracerebral hæmo-

rrhages, depressed fractures with laceration of the brain, and encapsulated meningeal hæmatomata. Such a clinical group do not amount to more than 6 per cent. of all infantile cerebral palsies. This monograph demonstrates clearly the present lack of accurate neurological investigation and classification based upon large series of cases of infantile spastic paralysis; secondly, it demonstrates the necessity for an investigation of the subsequent fate of a series of known non-fatal birth injuries, the obstetrical and early clinical data being available.

Part II. The second half of the volume deals with birth injuries of the spinal cord. The authors are to be congratulated in collecting a remarkable series of twenty-eight such cases. By the publication of these and analogous cases gleaned from the scattered literature, they have pointed the way to a veritable *terra incognita* of neurology.

Traction on the after-coming head has been the chief factor responsible for these injuries, which apparently vary not only in severity and distribution, but also in their ultimate outlook. Although no pathological evidence is forthcoming in the personal series of cases, it appears from the accounts of other published cases that all degrees of damage to the cord may occur—localized or diffuse—with or without lesions in the meninges or vertebral column, up to complete trans-section. The difficult task of physical examination has been carried out by the authors according to the "methods of the physiological laboratory," which are apparently synonymous with those of an incomplete neurological examination. In view of the peculiar value of the clinical cases it is unfortunate that the authors did not obtain the co-operation of a neurologist.

Convulsions et épilepsie des enfants. Par ANDRÉ COLLIN. Paris: Doin and Cie. 1926.

In the first section the author deals with the greater aptitude of the young towards the convulsive state, and three lines of argument are pursued—histological, chemical, and hormonic. Incomplete insulation of the immature neurone is comparable with instances of pathological sclerosis occurring in the adult—both conditions being attended by an exaggerated convulsant aptitude. Instability of the calcium metabolism of the infant is the second predisposing factor, the third being the incomplete development of the antitoxin function of the endocrine system.

A clinical discussion follows upon the epileptoid states in infancy; distinction is made between benign convulsions occurring in the offspring of neuropathic parents as a result of peripheral irritation or intestinal disorders, and the fits of essential epilepsy commencing at an unusually early age. The points of differential diagnosis and the dissimilar prognostic significance are then considered.

The diagnosis of idiopathic epilepsy arising in the first decade is carefully considered by the author.

A hypothetical consideration of the mechanism of epilepsy is undertaken and evidence is advanced to demonstrate a marked neurovegetative instability; such lack of equilibrium is regarded as the result rather than cause of cortical dysfunction. The prognosis of childhood epilepsy is markedly influenced, according to the author, by the onset of puberty; at this epoch many of the labile endocrine and metabolic factors become stabilized, often with permanent benefit to the convulsive state.

The monograph is concluded by a short chapter on treatment.

Les ramollissements sylviens. Par MAURICE-LÉVY. Paris: Doin et Cie. 1927.

The untimely death of Charles Foix struck a profound blow to the French neurology, for he was the instigator of a new type of clinico-pathological approach to the subject of cerebro-vascular disease. By his elaborate and painstaking correlation of clinical phenomena with the finer macroscopic anatomical data, a new neurological chapter was being compiled on the subject of neuro-angeiology. Fortunately, Foix had inspired a school of younger physicians who are continuing his work. The present monograph on the syndromes of the middle cerebral artery is a worthy contribution: it is an elaboration of a posthumous paper on the same subject by Foix and Levy, which recently appeared in the *Revue Neurologique*. The minute anatomy of the artery is first discussed and, in agreement with the work of Shellshear among British workers, it is shown that the branches are laid down with precision, and are constant in their distribution: abnormalities in the arterial arrangement consist in anomalies in the point of origin only: the ultimate destination is absolute. The course and distribution of the individual ramifications are described. Levy next deals with the symptomatology dependent upon occlusion of individual branches, and also of the main trunk at different levels. Thus one is lead up to the conception of complete and incomplete Sylvian softenings. The latter may be superficial or deep (partial and complete); or they may be of the nature of a partial or total "posterior" occlusion. Syndromes dependent upon lesions of individual arteries are next described, and the question of bilateral and multiple softenings is approached. Throughout the work the dictum of Foix is stressed, that in cases of partial or complete arterial obliteration the most ischæmic zone is that most proximal to the point of obstruction, while the more distal areas are relatively immune. The latter half of the book is occupied by records of illustrative cases—numbering forty-five in all—most of which occurred in the author's own practice.

Post-Encephalitic Respiratory Disorders. By SMITH ELY JELLIFFE. Nervous and Mental Disease Monograph Series, No. 45. New York and Washington. 1927.

The first part of this monograph is historical in scope and gives a valuable and detailed analysis of the numerous cases of respiratory disorder recorded to date. Full descriptions of the author's two cases follow, and details are given

of a psycho-analytical investigation. Case 1 lost all respiratory symptoms whilst under treatment; the outcome in Case 2 is not reported. The third and fourth sections of the work deal with phenomenology and psychopathological problems. The author has, in company with Runge and Witzel, emphasized the psychological factors at work in these patients and believes that the "encephalitic cases are doing the best they can with those pathways which are left open for functioning." This monograph is of interest, but contains much that will be inacceptable to present neurological thought. In places the argument is so vague as to be incomprehensible, and the writing savours more of mysticism than of medicine.

Les nouvelles méthodes sur les réactions colloidales du liquide céphalo-rachidien. Par EUGENE DE THURZO. Paris: Maloine. 1927. Pp. 180. Price 30 fr.

The first part of this monograph is occupied by a consideration of the author's modification of the collodial benzoin reaction. By the addition of "licht-grün" and fuchsin to the alcoholic solution of benzoin, a "bicolour" component is added to the ordinary precipitation test of Guillain. The author emphasizes the delicacy of the reaction in neurosyphilis. A colour modification of the colloidal mastic test is also detailed. A series of spinal fluid examinations was carried out, using Kafka's colloidal paraffin; the simplicity of this last test is commented upon, but a relative slowness in precipitation and the greater element of subjectivity in reading the tubes are points of disadvantage. The latter half of the book deals with the behaviour of the colloidal reactions in neurosyphilitic fluids after treatment, in disseminated sclerosis and encephalitis, and after artificial inflation of the ventricles with air. The varieties of colloidal shellac reactions are considered finally.

Les tumeurs de la moelle. Par J. A. CHAVANAY. Pp. 78. Paris: Doin et Cie. 1928.

This is a concise and interesting little résumé of the clinical manifestations of spinal cord tumours. The pathological and physiological aspects are not dealt with; the symptomatology is given in a clear and unequivocal manner, though many well-known features of a clinical nature are not mentioned. There is a chapter dealing with the spinal fluid changes, with the results of monometric studies, and with the injection of air or lipiodol into the subarachnoid space. The indications and results of surgical intervention are considered, and treatment by X-rays is referred to briefly. A short bibliography concludes the volume.

Anatomical, Phylogenetical and Clinical Studies on the Central Nervous System. By B. BROUWER. Baltimore: The Williams and Wilkins Co. 1927.

Within this volume are contained the three lectures delivered by Professor Brouwer at the University of Johns Hopkins. They set out in the author's

usual lucid and naive style the views upon many of the problems of neurology which are associated so closely with his name.

The first lecture deals with the projection of the retina on the brain: by means of a method of experimental ablation of special sectors of the retina, the secondary degenerations were traced by the Marchi method as far as the lateral geniculate body in a series of cats, rabbits, and monkeys. Brouwer's conclusions lend support mainly to the localization theories of Henschen; he finds that in the chiasma the fibres from the upper part of the retina cross dorsally, while the macular fibres lie chiefly in the middle. This clear-cut localization is recognizable, moreover, in the primary optic centres, and it is pointed out that as one passes from an oral to a caudal direction, the representation of the macula becomes relatively greater in area. Brouwer's conclusions concerning the cortical representation of the retina are not yet mature, but his impressions are that as regards the projection for binocular vision, the idea of an exact localization must be given the lead; furthermore, the projection of the macula must be localized, but nevertheless over a wide area—in this way supporting the view of Holmes. Throughout this work the author stresses the importance of localization in three dimensional planes.

Lecture 2 considers the pathology of sensibility: Brouwer approaches the subject from a biological aspect and deals with "gnostic" as opposed to "vital" sensibility. The former is subserved anatomically by the tracts in the posterior column, the mesial fillet, and the radiations between thalamus and post-central cortex. The pathways for transmission of vital sensibility are more complex, and the central representation is widespread and diffuse; the centripetal fibres of the automatic nervous system probably take part. By a series of comparative anatomical studies it is shown that the vital sensibility is the more primitive variety and must be localized to the antero-lateral columns: gnostic sensibility is the neotype, and does not appear in the phylogenetic scale until the amphibians, reptiles, or birds: it is of later development in the human fœtus. This form of sensation is localized to the posterior columns.

The last lecture concerns the significance of phylogenetic studies for the neurologist. The first example chosen is of a case of localized cerebellar atrophy occurring in an adult, and the light thrown by it upon the problem of cerebellar segmentation. Brouwer is led to agree in the main with Edinger's conception of neo- and palæo-cerebellum: the olivary system, too, appears as a duplicate structure, the accessory olives and the oral portion of the principal olive belonging to the palæo-cerebellar system, and the remainder to the neo-cerebellum. The author next takes the phenomenology of disseminated sclerosis as an illustration of diffuse lesions occurring haphazard throughout the central nervous system, but producing symptoms only in accordance with the principles of evolution and dissolution. The clinical results of such lesions are in the main the products of disorder of the newest and most highly evolved functions. Hence one finds abolition of the abdominal responses, rather than of the knee-jerks; inco-ordination and spasticity rather than paralysis; speech disorders before cranial nerve palsies; and nystagmus.

Idiosyncrasies. By Sir HUMPHRY ROLLESTON. London: Kegan Paul, Trench, Trübner and Co. 1927. Price 2s. 6d.

This little book, one of the Psyche Miniature Medical Series, is a most useful compendium of a subject which, though it is little understood, has a vast literature. The subject is discussed in the pleasantly worded, scholarly manner which one associates with the author.

The book is worth reading for the introduction alone; it would tax most people lucidly to define the term "idiosyncrasy." The sections on physiological hypersensitiveness, food and drug idiosyncrasies, and asthma, will be particularly useful, both to the practitioner and the student. Those who wish to study the subjects more extensively will find references to the literature quoted.

Psychologies of 1925. Edited by CARL MARCHISON. Clark University, Mass. 2nd Edition. Oxford: University Press. 1927. 28s. net.

In a comparatively brief space each of the outstanding figures in modern psychology gives a résumé of his point of view. The volume is divided into six parts, each devoted to a definite psychological approach.

Part 1 concerns "Behaviourism," and is written mainly by Dr. Watson, its protagonist, and by W. S. Hunter. In "What the Nursery says about our Instincts," and "The Experimental Studies on the Growth of the Emotions," Watson again propounds the physiological standpoint, which is tantamount to a complete denial of the autonomy of psychology, and the familiar psychological views of instinct.

In *Part* 2, Robert S. Woodworth explains his "Dynamic Psychology," which denies the validity of "Behaviourism," reasserts the agency of mind as a dynamic force, but does not leave the realm of biology, as many psychologists do.

Part 3 is devoted to the "Gestalt" psychology, and here its two great exponents, Koffka and Köhler, deal with mental development and the intelligence of apes respectively. The theory of "Gestalt" has little use for behavourism, although it accepts its experimental methods. Here mind reasserts itself fully, and the theory expounds, on the basis of considerable experimental research, the way in which the external world is apprehended as a system of configurations which mind imposes upon it.

Part 4, "The Purposive Groups" exposes the errors of behavourism, deals with the dynamics of co-consciousness and multiple personality, and a denial of the mechanical theory of man's mental life. These chapters are contributed by Morton Prince and McDougall.

Part 5 is concerned with "Reaction Psychology," by Knight Dunlap; and *Part* 6, "Psychologies called Structural," by Mavison Bentley.

Although there are striking divergencies of view in this volume of surveys, one welcome feature is outstanding and admitted by all writers, that is, the insistence on biological categories in the treatment of all psychological problems.

Zwang und Zweifel. Teil 1. By WILHELM STEKEL. Wien: Urban und Schwarzenberg. P. 633. 1927. Price M. 30.

This volume is the first part of the ninth volume of Stekel's collected works: "Störungen des Trieb und Affektlebens."

In this enormous volume he deals with the nature and psychotherapy of Compulsion and Doubts. He classifies compulsive neuroses according as they arise from some anxiety about the organs (principally sexual), as result of education, and lastly because of the Ego demands. He distinguishes psychopathic doubt from philosophic doubt or scepticism; for him it is merely a symptom of deeper affective disturbances. The book is more or less free of any systematic attack on psychological problems, and from his copious case-histories and his discoveries, it is obvious that he manages to achieve understanding and cure without reference to the Unconscious or any other Freudian mechanism. But he never hesitates to take the cream of other people's discoveries and apply them with a hearty optimism, which is very amusing and frequently instructive.

Das autistisch-undisziplinierte Denken in der Medizin und seine Überwindung. By E. BLEULER. Berlin: Julius Springer. 1927. 4th Edition. Price M. 7.50.

This edition differs from its predecessors only in the addition of some important footnotes to the text. The book deals with the general character of Autistic Thinking, that is, that type of mental activity which goes on independent of reference to external reality. To this extent it is undisciplined, i.e., ungoverned by the logical demands of external reality It must, Bleuler says, be sharply distinguished from "egoistic" thinking which is subject to the discipline of the organized Ego.

The volume is full of interest not only to the psychiatrist but for the general physician. The book is illuminated throughout by that calm wisdom which has always been expected and given in the writings of Bleuler, perhaps the sanest of living psychiatrists.

Études sur la circulation de l'influx nerveux dans l'arc réflexe. By A. RADOVICI. Préface de G. BOURGUIGNON. Paris: Masson et Cie. Price 16 francs.

Dr. Radovici's monograph is an experimental investigation of the application of the chronaxic theory of Lapicque to human nerve physiology. It demonstrates the possibility of extending the theory from neuro-muscular phenomena, in particular as worked out by Lapicque and Bourguignon, to intraneural excitation of the axon itself. His researches establish definitely the rôle of the "time" factor in all laws governing the relationship between stimulus and response. In experiments made on man with the cord "released" by compression lesions, it is demonstrated that the automatic and rhythmic

reflex movements in the limbs below the lesion have a frequency varying according to a definite law, corresponding also with the frequency of the electrical stimulation. The law is identical with Weber's law of sensation, Pfeiffer's tropism phenomena, and with Hedwig's observations on embryonic development. His observations are also found to agree with Lehman's work on the retina, and Waller's on the optic nerve.

Weber's law of the relation between stimulus and sensation, in other words, is nothing else than a purely neurological law, and psycho-physical parallelism becomes in consequence electro-physical parallelism.

Beiträge zur Kenntnis der mongoloiden Missbildung. By M. W. M. van der SCHEER. Berlin: S. Karger. 1927. Price M. 12.

This monograph is a statistical, anatomical and clinical investigation of the part played by the uterine mucous membrane and the amnion in the production of the deformity known as mongolism.

The study is a very thorough one and the subject of definition of terms is carefully considered. The first section deals with general considerations and the anthropological arguments are discussed: and an interesting compromise is suggested between Crookshank's purely racial explanation and the embryological ætiology of the condition. It is the latter which concerns the author, and he deals statistically and clinically with the affections of the uterine mucosa which may cause the deformities. The third section discusses the specific somatic and psychical features of mongolism.

The monograph is useful because all ætiological factors are considered and the differential diagnosis from mental deficiency is constantly being borne in mind.

The Psychology of Murder. A Study in Criminal Psychology. By A. BJERRE. Longmans, Green and Co., Ltd. 1927. 9s. net.

Bjerre occupied an important position in the Danish judicature and had ample opportunity of studying the details of each of the cases with which he deals. Although he is of the opinion that the cases he had under investigation were capable of classification under three heads: self-deception, anguished fear, and shamming, he has noted common qualities binding all clinical manifestations together, namely, psychological emptiness and a tendency towards general mental weakness. In each case a very full account has been given of the crime which brought each delinquent to justice, and the background of the crime is carefully looked into. However this may be, the author treats throughout of the superficial mechanisms that actuated the crime, and of the essential weakness exhibited in facing environment, and in so concentrating his attention he has lost the inwardness of the whole criminal psychology. The unconscious promptings of crime, the deep compensatory mechanisms, are missed and are merely dealt with as if the compensations were done deliberately

in order to gain easier ends. These criticisms while diminishing the scientific value of the book do not detract from its practical use. The book remains an interesting contribution to criminology, particularly as the author opens up the avenue to the pathological approach as against the legal approach to the study of crime.

Selected Papers on Psycho-Analysis. By KARL ABRAHAMS. International Library of Psycho-Analysis. No. 13. London: Hogarth Press. 1927. Price 30s.

This volume is in the nature of a memorial to the late Karl Abrahams, who was perhaps, after Freud, the outstanding pioneer of psycho-analysis. It is a collection of all the writings he contributed on the subject, and illustrates the many-sidedness of his work, and this renders it impossible in a short review to deal adequately with the mine of information he has worked so assiduously. Firstly, it illustrates his pre-occupation with the evolution of the child-mind, and the contribution that the affective life of the child makes to adult psychology. Secondly, it shows his interest in the psychoses and the light that psycho-analysis attempts to throw on their development. Thirdly, his wide cultured interests and linguistic powers are manifest in his researches into applied psychology, that is, psycho-analysis applied to folklore, history and education.

Dr. Ernest Jones gives an interesting memoir of the author and schematizes his positive contribution to the subject and his application of Freud's doctrines.

(1) *Correlated Physiological and Morphological Studies of the Development of Electrically Responsive Areas in the Cerebral Cortex of the Opossum.* (2) *Histological Development of Cerebral Motor Areas in Young Kittens correlated with their Physiological Reaction to Electrical Stimulation.* By ORTHELLO R. LANGWORTHY. "Contributions to Embryology," Nos. 103 and 104. Reprinted from Vol. xix of "Contributions to Embryology." Carnegie Institute, Washington. Publication No. 380. 1927. Pp. 149 and 177.

In these studies the author has used the pouch-young of the opossum and newly-born kittens in order to determine the order in which the motor areas for the various segments of the body become differentiated and electrically excitable. By using the pouch-young of Virginian opossums he obtained what were practically air-breathing embryos, as these animals are born when only 11 mm. in length. They appear to climb up into the pouch by grasping the hair with their fore-limbs, and in accordance with this the author found that the fore-limb centre was the first to become differentiated and electrically excitable. No area which produced movements of the hind-limbs was discovered by him, even in adult opossums. In this his findings agree with those

of Cunningham. In the kitten also the area for the fore-limb was excitable at birth, and this was followed by the area for the hind-limb at 16 days, and the area for the face at 21 days. These dates corresponded with the state of maturity of the cortex and not with the myelination of the pyramidal tract, as this is not myelinated at all till the age of 9 or 10 days.

Certain other points, such as the development of Brodmann's *area frontalis agranularis*, were also brought out by these experiments.

Zellaufbau der Grosshirnrinde des Menschen. By Dr. CONSTANTIN VON ECONOMO. 8. 146. Berlin: Julius Springer. 1927. Price 18 marks.

L'architecture cellulaire normale de l'écorce cérébrale. French edition, translated by Dr. LUDO VAN BOGAERT. Pp. 183. Paris: Masson et Cie. 1927. Price 80 francs.

The author prefaces this work by the explanation that it is not meant to replace his large atlas, but rather to form an introduction to it. In any case it is of itself a very useful book, as it presents the detailed cell structure of the cerebral cortex in an easily assimilable form. The 109 types of cortex are grouped into five main forms which are probably functionally as well as histologically distinct. By his system of naming each of the cortical areas by a letter-group, the author preserves the gross anatomical nomenclature, and at the same time gives an indication of the type of cortex present in each area. This system seems to us a definite improvement on the numerical system of Brodmann, although, whether as a result of it, or for other reasons, the number of different cortical areas is more than doubled.

The book is beautifully illustrated, and the forty-six large photomicrographs of the most distinctive cortical areas make it a book which will be as useful for reference in the laboratory as for home study. The French translation by Dr. Ludo van Bogaert has been well done, and the addition in it of headlines to the paragraphs in leaded type facilitates reference. Both volumes are most welcome additions to the neurologist's library.

Embryology of the Neuromuscular Spindle. By FIDEL CUAJUNCO. "Contributions to Embryology," No. 99. Carnegie Institute, Washington. Publication No. 380. 1927.

This small monograph is a record of an investigation into the development of the neuromuscular spindle and of its nerve-supply in pig embryos. The author confirms the classical researches of Sherrington and of Batten in finding the spindles supplied with sensory and not with motor nerve-endings, but he also found some twigs which appeared to be of sympathetic origin terminating within the capsule. It is interesting to note that the nerve-supply reaches the embryonic muscle fibres before they become segregated off by the capsule of the spindle.

The illustrations are excellent, but in those of the complex nerve-endings it is not always clear how much is photomicrograph and how much drawing. Undoubtedly good drawings illustrate the author's intentions better than the best photomicrographs, and a little touching up of photographs is excusable, but this does not altogether exculpate the author from describing as a photomicrograph what appears to be, as far as the more important details are concerned, the result of pen and ink.

Die Lagereflexe beim Menschen. By HANS HOFF und PAUL SCHILDER. S. 182. Wien: J. Springer. 1927. Price 12 marks.

The authors have defined their book as an account of the attempt to apply to the clinical material at the Vienna clinic those principles of posture regulation laid down by Magnus in his work on "Körperstellung." It contains firstly a very complete and up-to-date summary of the reflexes worked out in the school at Utrecht. Following this, there is a further summary of the anatomy of those tracts and central nervous mechanisms, particularly of the red nucleus and its connections, which are held to supply the machinery for these reactions.

The main body of their book is concerned with observations on the postural reflexes in adults, but is preceded by a shorter account of those already described in the normal infant by Minkowski, Gamper, Schaltenbrand and others, and their contribution to this part of the subject consists in a very useful résumé of the appearance and disappearance of these reflexes as age advances.

With those reflexes which appear in premature infants, or those with various forms of cerebral agenesis or trauma, they are less concerned, although these form an interesting confirmatory link between the experimental animal and the normal child.

It appears from these observations that the transition of reflex activity from the condition of the infant at birth to that of the normal adult may be divided conveniently into four periods. The first lasts from birth to 6 months, during which the reactions to movement (Bewegungsreacktionen) are prominent; the next a quadruplegic condition, in which the characteristic primitive postural reflexes, as described in animals, are to be seen; the third, in which the tonic influences which determine the quadruplegic position gradually diminish, with the assumption of the erect attitude, and the final stage, complete by the fifth year, in which the postural reflexes have assumed a form which they maintain throughout adult life.

The authors have elaborated a series of clinical tests which are clearly and practically detailed, and of which the normal variations in response are precisely given. The results of the application of these tests to patients suffering from a variety of nervous lesions, especially lesions of the cerebellum, provide the material for most of the rest of the book which closes with a tabular summary of the chief results therein detailed.

It is clear that many reactions in man find no counterpart in the behaviour of experimental animals, and that only by analogy can their mechanism be localized with any degree of probability within the mid-brain. Many of these have, however, been shown to yield definite localizing value as clinical tests on purely empirical grounds.

The authors have taken great care in their attempt to decide how far the defect which accounts for the appearance of such reactions is sensory or motor in nature, and herein lies the chief basis for criticism of this work, in that, as they would readily themselves admit, this discrimination is seldom possible with certainty, and indeed, in some cases the defect may be in part neither sensory nor motor, but definitely perceptual as in the Lagebeharrungsversuch and the Imitationsphänomen.

This detracts in no wise from the very considerable value of the book as a whole, and the detailed bibliography at the end is admirably complete and clear.

Les réflexes de posture élémentaires. Par P. Delmas Marsalet. Pp. 176. Paris: Masson et Cie. 1927. Price 16 francs.

In this book Marsalet has recounted his investigations of the clinical occurrence described by Foix and Thevenard, which consists in the shortening of a muscle secondary to its passive relaxation, such shortening being, in their opinion, a response on the part of the muscle designed to fix it "without slack" at the newly-imposed length. Working with the tibialis anticus muscle, the procedure of Delmas Marsalet has been to approximate its origin and insertion passively and quickly by sudden dorsiflexion and inversion of the ankle, and by means of a tambour fixed over the subcutaneous portion of its tendon, where it bridges over the ankle, to record the subsequent readjustment of tension by active contraction of the muscle.

He has found abolition of this reflex in polyneuritis, tabes, flaccid hemiplegia and cerebellar hypotonia; its replacement by an intermittent clonic response, in cases of damage of the pyramidal tract, and its exaggeration in cases of Parkinsonism. He then proceeds to describe the various factors which can be used to modify at will the reflex examined; continued passive movements of the limb which transitorily diminish the reflex and various drugs, the most prominent of which was scopolamine, which abolish it when in sufficient (1·25 mgm.) doses after an interval of three-quarters of an hour.

He has come to the conclusion that the reflex is a widespread one, involving many nervous pathways, and that its nervous mechanism is quite other than that of the tendon reflexes. He finds further that in cases of Parkinsonism there is a strict correspondence between the distribution of the rigidity and the occurrence of exaggeration of the postural reflexes—a correspondence which is both topographical and proportional in degree, so that he is led to ascribe a causal relationship to the two phenomena, and states that all the characteristic features of this condition dependent upon rigidity are due to the exaggeration of the postural reflexes.

Unfortunately there is no serious attempt to substantiate the hypothesis that this phenomena is reflex and no evidence that the explanation offered is the correct or even the most probable one.

The interpretation of this reaction in the terms of the physiology of nervous reflexes is seriously open to question, and it is a pity that this type of response should be described as a postural reflex when this term has been already used to describe the series of delicately adapted reaction so ably and scientifically investigated by Magnus and his workers, and to which this bears no relation whatever.

J. Hughlings Jackson: *Ueber Aufbau und Abbau des Nervensystems.* Uebersetzt und eingeleitet von Otto Sittig. S. 137. Berlin: S. Karger. 1927. Price 6.60 marks.

Sittig's translation into German of Hughlings Jackson's "A Study of Convulsions," has been already noticed in this Journal. He has now translated Jackson's "Croonian Lectures," which is one of his most important articles, and contains some of the most valuable contributions he made to the study of the nervous system in health and disease.

As on the previous occasion, the English text and the German translation appear on opposite pages; the value of this to everyone to whom English is a foreign tongue is obvious, and fortunately it also makes available to English readers these lectures which have never been reprinted since they originally appeared in the *Lancet*. The translation seems to be accurate and readable.

Epidemic Encephalitis. By L. M. Crafts. Pp. 237. Boston: R. E. Badger. 1927.

This volume is based on the author's own experience, but it also contains references to the literature of epidemic encephalitis. It presents concisely the main clinical features of this polymorphous disease, and discusses its differential diagnosis in considerable detail. Half its pages are devoted to case reports, most of which will be of use only to the student unfamiliar with its ordinary symptoms.

Kompendium der topischen Gehirn und Rückenmarksdiagnostik. Von R. Bing. 7te Auflage. S. 259. Berlin: Urban und Schwarzenberg. 1927. Price 10 marks.

The successive editions of Bing's well-known textbook on the diagnosis of local lesions of the nervous system have been noticed in this Journal. The seventh edition has grown but little in bulk, but both the text and illustrations have been revised. The chief alterations have been made in the sections on the cerebellum, the basal ganglia and the visual system, but fuller reference will be found here to the postural reflexes and spinal automatism, subjects in which more exact knowledge has been recently acquired. The book remains one of the best of its scope available to the student of organic neurology.

Der Tonus der Skelettmuskulatur. Von E. A. SPIEGEL. 2te Auflage. S. 203. Berlin: J. Springer. 1027. Price 18 marks.

The first edition of this monograph, which appeared under the title, "Zur Physiologie und Pathologie des Skelettmuskeltonus," has been already reviewed (*Brain*, 1924, **47**, 257). This, the second edition, is almost double its size, and contains much new matter as well as amplifications of many of the original sections. The further development has been chiefly in the direction of investigation of the nature muscle tone by animal experiments, and little reference is made to the clinical applications of the results obtained. The chief conclusions are that: (1) In the labyrinthine nuclei and in the formatio reticularis of the hind-brain are supraspinal centres for extensor or antigravity tone; (2) the red nucleus is a centre which acts by inhibiting extensor tone, or by regulating the distribution of tone between the flexors and extensors; (3) the basal ganglia contains a reflex apparatus which can modify the relative tone of agonists and antagonists; (4) the cortico-spinal and cortico-pontine systems tend to inhibit extensor tone; (5) the cerebellum is a proprioceptive reflex mechanism which, on the one hand, acts as a brake, on the other diminishes the tone of the extensors and increases that of the flexors. (6) The autonomic nervous system has no influence on tone. These conclusions serve to indicate the importance of this monograph for all interested in the question of muscle tone, a subject that is daily taking a larger part in clinical neurology.

Mikroskopisch-topographischer Atlas des menschlichen Zentralnervensystems mit begleitendem Texte. Von O. MARBURG. 3te Auflage. S. 226. With 40 Illustrations in Text and 32 Tables. Wien: F. Deutiche. 1927. Price 28 marks.

It is now twenty-five years since the first edition of Marburg's Atlas appeared, and nearly twenty years since the second was published. The third edition is considerably enlarged, especially the descriptive pages and the sections on the cellular architecture of the cerebral cortex, and many new illustrations are provided. The Atlas still retains its essential features; a large number of excellent and wonderfully reproduced drawings of sections in various planes of all levels of the central nervous system are given, and the text consists mainly in descriptions of the individual sections. This method of study of anatomy is perhaps essential for the beginner, but alone it unfortunately fails to present to the student the clear picture of the structural connections of different parts of the brain and cord which is required in the practice of neurology. All the illustrations are fully legended, and a table gives the name in German, French, English and Italian of each structure indicated. In this table the figure in which tract or nucleus can be seen is also shown, so that anyone who is interested in any structure, as for instance, the acoustic radiation, can at once find its position and trace its course in the illustrations.

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EDITOR.